Vascular lesions of the gastrointestinal tract

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Abstract

Vascular lesions of the gastrointestinal (GI) tract include arterio-venous malformations as angiodysplasia and Dieulafoy's lesion, venous ectasias (multiple phlebectasias and haemorroids), teleangiectasias which can be associated with hereditary hemorrhagic teleangiectasia (HHT), Turner's syndrome and systemic sclerosis, haemangioma' s, angiosarcoma' s and disorders of connective tissue affecting blood vessels as pseudoxanthoma elasticum and Ehlers-Danlos's disease.

As a group, they are relatively rare lesions that however may be a major source of upper and lower gastrointestinal bleeding.

Clinical presentation is variable, ranging from asymptomatic cases over iron deficiency anaemia to acute or recurrent bleeding that may be life-threatening. Furthermore, patients may present with other symptoms, e.g. pain, dysphagia, odynophagia, the presence of a palpable mass, intussusception, obstruction, haemodynamic problems resulting from high cardiac output, lymphatic abnormalities with protein loosing enteropathy and ascites, or dermatological and somatic features in syndromal cases. Diagnosis can usually be made using endoscopy , sometimes with additional biopsy. Barium radiography, angiography, intraoperative enteroscopy, tagged red blood cell scan, CT-scan and MRI-scan may offer additional information. Treatment can be symptomatic, including iron supplements and transfusion therapy or causal, including therapeutic endoscopy (laser, electrocautery, heater probe or injection sclerotherapy), therapeutic angiography and surgery. The mode of treatment is of course depending on the mode of presentation and other factors such as associated disorders. If endoscopic or angiographic therapy is impossible and surgical intervention not indicated, pharmacological therapy may be warranted. Good results have been reported with different drugs, albeit most of them have not been tested in large trials. (Acta gastroenterol. belg., **2002**, 65, **213-219**).

Key words : gastrointestinal (GI) tract, vascular, malformation, phlebectasia, telenangiectasia, haemangioma, angiosarcoma, connective tissue disease.

Introduction

Vascular lesions of the gastrointestinal tract as a group are relatively rare lesions, with an annual incidence of 1 in 14000 individuals, as reported in 1 large series (1). Nevertheless, they are thought to be responsible for 2-5% of all episodes of acute upper gastrointestinal bleeding and up to 35% of cases of lower gastrointestinal bleeding. Clinical presentation however is variable.

Several authors have proposed a classification system for this disorders. The system most commonly used is based on the type of vessel that is affected combined with dermatological or somatic features, as proposed by Camilleri *et al.* (1). This classification is presented in table 1.

Each of these lesions will be discussed further in detail.

I. Angiodysplasia

Angiodysplasia is one of the most common causes of lower gastrointestinal tract bleeding in the elderly (over 60y), together with diverticulosis. The prevalence in asymptomatic people is estimated around 0,83% and the incidences in asymptomatic and symptomatic patients are estimated around 3 and 6-12% respectively. Symptomatic patients may present with melaena, haematoschezia, iron deficiency anaemia and rarely massive haemorrhage (1-3). Up to 80% of the lesions occur in the right colon. They are thought to be degenerative in origin, resulting from chronic intermittent obstruction, leading to dilatation of veins, venules and capillaries with subsequent loss of competence of the precapillary sphincter, ultimately resulting in an arteriovenous communication. Rarely, other parts of the gastrointestinal tract as the small bowel may be affected. In these cases patients are usually younger than 50 years and lesions might be congenital (1,2,4). Some have reported an association between aortic stenosis, atherosclerosis, chronic obstructive pulmonary disease, cirrhosis and collagen vascular disease on the one hand and angiodysplasia on the other hand, but this has been doubted by others on the grounds of methodological deficiencies in some large studies (5,6). Furthermore, there have been several reports on a link between angiodysplasia and von Willebrand's disease. This association might not be causal but just represent an increase in symptomatic cases due to a higher propensity to bleed. Moreover, as stated by Warkentin *et al.*, any vascular disease may lead to clearance of larger multimer forms of von Willebrand factor (vWf). Indeed, vascular disease may cause high shear rates that induce platelet adhesion which in turn is dependent on interaction of platelet membrane glycoproteins with vWf. This may lead to overconsumption of vWf multimers that might be needed to prevent bleeding in angiodysplastic lesions. Increased bleeding in uremic patients can also be explained in this way because uraemia is associated with disturbed vWf dependent haemostasis (7,8).

Diagnosis of angiodysplasia is usually made on endoscopy that might reveal a flat, cherry-red lesion, 2-10 millimetres in diameter, with a raised central 'feeding' vessel and a flare of radiating peripheral vessels,

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Table 1. — **Classification of lesions the gastrointestinal tract (modified after Camilleri *et al.*) (1)**

I	Arteriovenous malformations * angiodyplasia * vascular ectasia Dieulafoy's lesion
II	venous ectasia * multiple phlebectasia * hemorrhoids
III	teleangiectasia * hereditary hemorrhagic telangiectasia (Weber-Rendu-Osler's disease) * Turner's syndrome * CREST and systemic sclerosis
IV	hemangioma * capillary * cavernous * mixed * Peutz-Jegher's syndrome * blue rubber bleb nevus syndrome * Klippel-Trenaunay-Weber syndrome
V	angiosarcoma
VI	disorders of connective tissue affecting blood vessels * pseudoxanthoma elasticum * Ehlers-Danlos's syndrome

resulting in a spidery appearance. On mesenteric angiography, angiodysplastic lesions present as tortuous, slowly emptying veins, vascular tufts or early filling veins. If these investigations are negative in a bleeding patient, a nuclear red blood cell scan can be performed (1,2,4,5).

When biopsied, angiodysplastic lesions present as a submucosal cluster of dilated arterioles, veins, venules and capillaries. Most of them are thin walled and have no muscle layer (fig. 1). In a later phase, the lesion might extend into the mucosa that may be thinned and ulcerated (1,2,5).

As stated in the introduction, treatment depends on severity of symptoms and comorbidity and includes wait and see, haemodynamical stabilising, endoscopic ablation and therapeutic angiography with embolisation or vasopressin injection. Also pharmacological treatment with somatostatin analogue and oestrogen have been used (1-5,7).

II. Venous ectasias

Multiple phlebectasias are non-neoplastic venous varicosities with a normal endothelial lining, not associated with somatic or dermatological features. Presumably they are congenital in origin but most often present in the middle-aged and elderly with signs of chronic or acute bleeding, that very occasionally can be life-threatening. The lesions most often are located in the submusosa, with thinning of the overlying mucosa. They are much less common than angiodysplasia and most commonly located in the mid-jejunum. They might also occur in the oesophagus and rectum. They often are very difficult to visualise on angiography and diagnosis often is delayed until laparotomy. Surgical intervention is the treatment of choice in symptomatic cases. Increased use of enteroscopy has identified the lesions as multiple dark or bluish red compressible nodules with a diameter ranging from a few millimetres to several centimetres (1, 4).

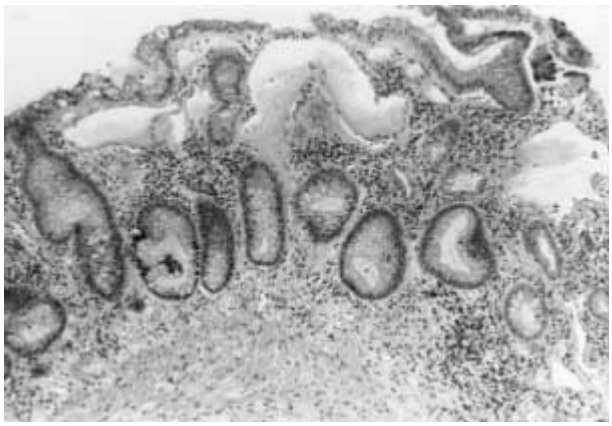

Fig. 1. — Angiodysplasia : presence of dilated capillaries in the colonic mucosa. Orginial magnificatio × 100.

Haemorrhoids are well-known recto-anal venous ectasias with a prevalence of up to 76%. They are responsible for 2-9% of acute lower gastrointestinal bleeding (5).

III. Teleangiectasias

Teleangiectasias are localised dilatations of capillaries and venules, and can occur sporadically or in a syndromal setting.

Hereditary hemorrhagic teleangiectasia (HHT) or Rendu-Osler-Weber's disease is an autosomal dominant disorder with an age dependent penetrance which is nearly complete by the age of 40-45. According to different reports, it has a prevalence of 1 in 2531 to 1 in 39000 (1,9). HHT is caused by a germ line mutation in the genes for endoglin or ALK, which both are members of the TGF-beta receptor family of proteins that are primarily expressed on the surface of endothelial cells. This probably results in disturbed migration and proliferation of endothelial cells, together with deficient matrix production and organisation. This interferes with the normal repair of the vessel wall, resulting in a cascade of effects and ultimately dilatation of post capillary venules. Eventually, the dilated venules connect to enlarging arterioles through capillary segments that later disappear, thus creating an arteriovenous connection (4,9). HHT might be associated with von Willebrand's disease and a hemostatic defect – hence the name – which may render treatment difficult (1).

Teleangiectasias can be found on the skin (most often on the lips, face and around ear lobes), lung, brain, liver, genito-urinary tract and throughout the gastrointestinal

tract. Mucocutaneous lesions, gradually increasing in number, usually appear in the second and third decades. The first symptom however often is epistaxis in childhood. Chronic gastrointestinal bleeding commonly starts in the forth decade and is present in 10-20% of patients (1,9,10). In some affected families, there also is involvement of lung, liver and brain with formation of arteriovenous malformations with high volume shunting that may lead to hypoxemia, systemic embolisation, high output cardiac failure and brain abscesses (4).

The diagnosis can be made on endoscopy, red blood cell scan and angiography but it can be very difficult to localise the bleeding vessel.

Therapeutic endoscopy is often the treatment of choice. Pharmacological treatment with oestrogens has been reported but is not well documented. Surgical intervention is often difficult due to the multifocality of the lesions. Embolisation has been performed for lung arteriovenous malformation and orthotopic liver transplantation for major symptomatic liver involvement (4).

Turner's syndrome is a well-known genetic disorder with a 45XO karyotype, resulting in a number of somatic features (webbing of the neck, short stature, coarctation of the aorta, renal abnormalities and lymphoedema, ovarian dysgenesis and congenital malformations of other internal organs). In Turner's syndrome, telangiectasias may occur throughout the gastrointestinal tract, most commonly in the small intestine. Haemorrhage resulting from these lesions is usually intermittent , self-limiting and very occasionally massive. In the latter case, surgical intervention can be needed but most often a conservative approach is warranted because the lesions regress with age (1,4).

Systemic sclerosis and CREST syndrome (calcinosis, raynaud phenomenon, oesophageal dysfunction, sclerodactyly and teleangiectasia) are related disorders with an annual incidence of 1 in a million. Genetic and environmental factors lead to an increased deposition of collagen but the exact mechanism is unclear (11,12). Because of the similarities between both diseases, Akesson *et al.* propose the term limited cutaneous systemic sclerosis in stead of CREST (13). In a series of Duchini *et al.*, 15,2% of patients had 1 or more episodes of gastrointestinal bleeding, about 40 % of which were caused by mucosal telangiectasias, that may occur anywhere in the gastrointestinal tract. Furthermore, there seems to be an association with GAVE (1,4,12,13). Histological examination shows non-specific alterations, i.e. dilated capillaries with some perivascular infiltrates and sometimes evidence of leakage (12).

Recognition of the clinical features should alert the clinician. Intestinal lesions can be identified and treated on endoscopy. Pharmacological treatment using oestrogen-progesterone preparations, desmopressin and aminocaproic acid has also been reported. In very severe cases, surgical intervention can be considered (4,12).

IV. Dieulafoy's lesion

Dieulafoy's lesion is a vascular malformation with unknown incidence because of many asymptomatic cases. It might be responsible for 1-2% of cases of upper gastrointestinal bleeding and can occur on any age. The lesion most commonly occurs in the most proximal 6 centimetres of the stomach but occasionally other localisations as the lower gastrointestinal tract are encountered. In the stomach, the lesion is caused by an artery that directly derives from the left gastric artery, without an intervening branching submucosal plexus. The large, pulsating submucosal artery may render the overlying mucosa weaker which may result in ulceration. Patients may thus present with often massive and repeated haematemesis, melaena, haematoschezia and hypotension or shock. A relation with NSAID's has been suggested but has not been proven (4,14-16).

Diagnosis can be very difficult because of the small size of the lesion. Repeated endoscopy might be needed to reveal a small protruding vessel that , lying in a small defect or covered with a cloth. Angiography might reveal the bleeding point but sometimes surgical exploration with gastrotomy is warranted (14-16).

Treatment again depends on the mode of presentation and includes haemodynamical stabilisation and interventional options as therapeutic endoscopy and angiography with embolisation. In about 5% of cases, surgical segmental ectomy is needed, after localisation of the bleeding vessel (4,14,16).

Careful histological examination of the resected specimen may show alterations that reflect the pathophysiology : a large (1-3 mm) but otherwise normal submucosal artery, often with a tortuous course can be found in the proximal stomach, most often at the lesser curve. There is no evidence of vasculitis, aneurysm, atherosclerosis or surrounding inflammation. The overlying mucosa might show a minute ulcer, through which the vessel might protrude (fig. 2) (14-16).

V. Gastric antral vascular ectasia (gave) – the watermelon stomach

Gastric antral vascular ectasia (GAVE) is a rare vascular disorder, occurring in the elderly, especially women. Patients present with features of persistent or chronic bleeding that might require transfusion (17,18). The lesion is identified on endoscopy as 'longitudinal rugal folds, transversing the antrum and converging on the pylorus, each containing a visible convoluted or sacculated column of vessels, the aggregate resembling the stripes of a watermelon', as originally described by Jabbari *et al.* The ridges may be raised, flat, mixed or there may be diffuse and scattered lesions, with also upper gastric involvement. Sometimes, the ectatic vessels do not present with a watermelon-like appearance but rather as aggregates of red spots. In this case the term gastric vascular ectasia (GVE) is used. This lesion

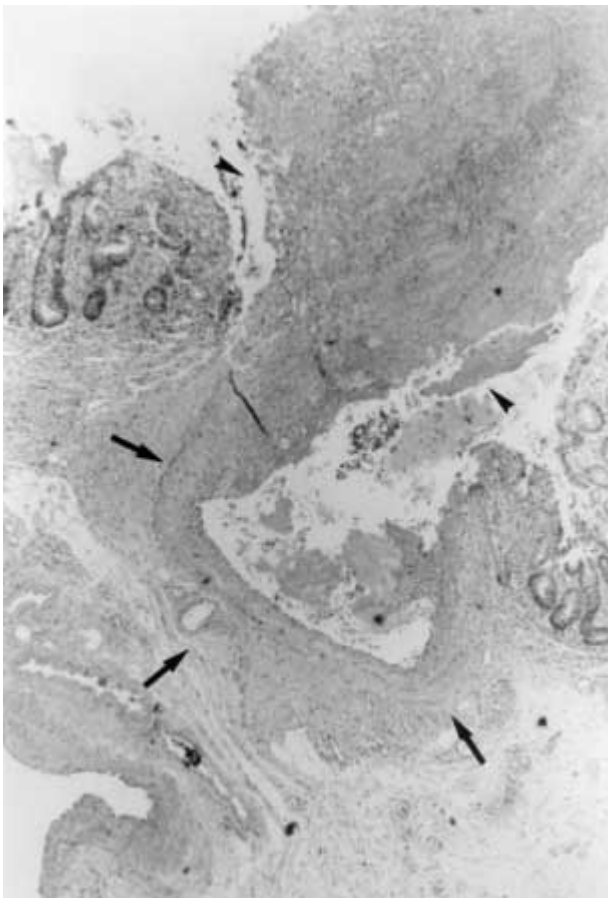

Fig. 2. — Dieulafoy's lesion : An unusual large artery (arrows) is present in the submucosa of the colon. The overlying mucosa is eroded, a process which also involves the artery (arrowheads).

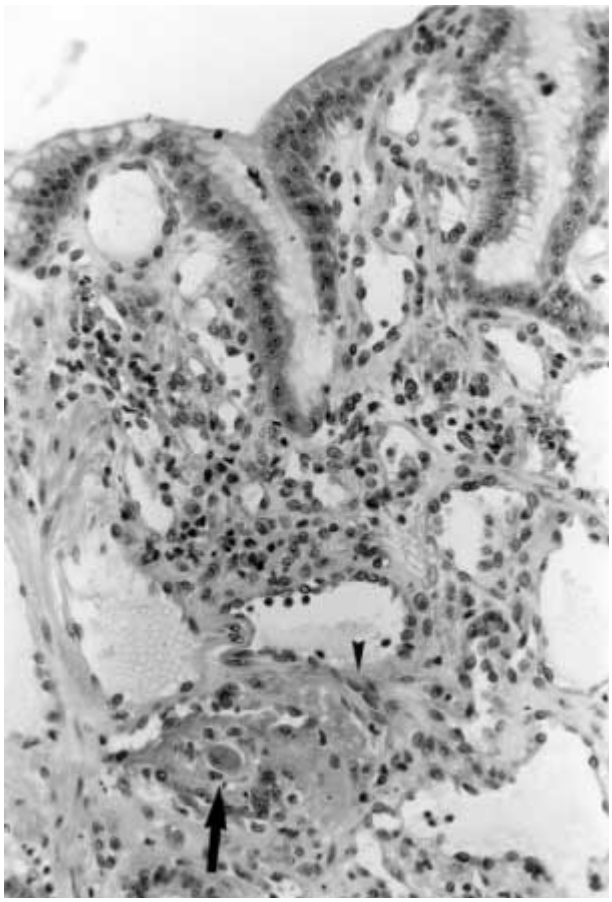

Fig. 3. — GAVE (Watermelon Stomach) : Presence of dilated capillaries in the antral mucosa, with focal thrombosis (arrow) and discrete myofibroblastic proliferation(arrowhead). Original magnification × 400.

must be differentiated from portal hypertensive gastropathy (PHG) which in severe forms also may give rise to aggregates of red spots. However, these spots in GVE always occur in association with a mosaic pattern or scarlatina rash appearance of the background mucosa. Endoscopic ultrasound might be useful in cases of doubt (17,19) (20,21). The diagnosis of GVE can be confirmed by histological examination of pinch biopsies. The mucosa is composed of hyperplastic glands, surrounded by a lamina propria with mild chronic inflammation. There are several dilated vessels, which often contain microthrombi and there is proliferation of myofibroblasts. The remainder of the mucosa may be normal or atrophic (fig. 3). In resected specimens, there is also oedema in the submucosa where also tortuous vessels can be noted.

Dilated vessels can also be encountered in PHG (fig. 4) but the presence of microthrombi and the proliferation of myofibroblasts favor a diagnosis of GVE in cases of doubt. Establishing a correct diagnosis is important because most PHG patients respond well to transjugular intrahepatic portosystemic shunts (TIPS), whereas TIPS should be avoided in GVE patients (17, 19-22).

The exact mechanism that causes GAVE is still unknown. It is thought to result from antral mucosal prolapse through the pylorus, caused by vigorous antral contractions, with consequent elongation and dilatation of mucosal vessels. This is partly confirmed by the fibromuscular proliferation that can also be seen in other prolapse diseases as haemorrhoids, stoma and solitary rectal ulcer. Other suggestions are a response to hypergastrinemia (because of the association with hypergastrinemia) or other vasoactive hormones as vasoactive intestinal peptide (VIP) or 5-hydroxytryptamine.

Treatment depends on the rate of blood loss. Therapeutic endoscopy has yielded excellent results. Furthermore, pharmacological therapies with prednisone, prednisolone, oestrogen-progesterone preparations, interferon alpha and serotonin antagonists have shown promising results but their efficacy has not yet been tested in large trials. In some cases, surgical intervention with Billroth I resection might be needed (17-19).

VI. Hemangioma

Haemangioma' s are tumors that are predominantly composed of blood vessels. Most of them are in fact

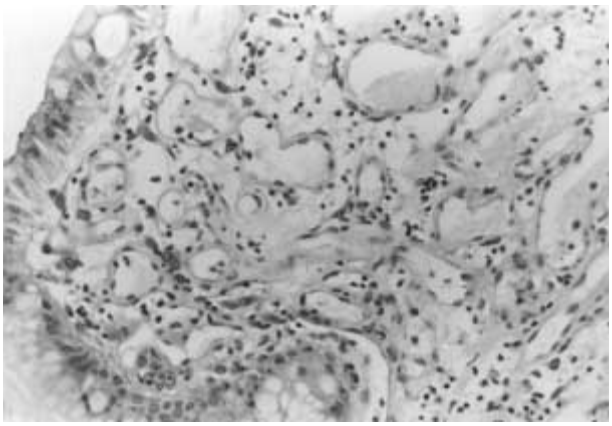

Fig. 4. — Hypertensive gastropathy : Presence of numerous dilated capillaries in the antral mucosa, without evidence of thrombosis of myofibroblastic proliferation. Original magnification × 400.

hamartomas. They are thought to represent 10% of all benign intestinal tumours but overall their presence in the gastrointestinal tract is rare : in '95 around 200 cases had been described in the small intestine and oesophageal haemangioma was present in 0,015% of patients in a series of nearly 20000 autopsies (4,23,24).

Haemangioma' s may be solitary or multiple. Some cases are asymptomatic, others present with a mass effect (pain, odynophagia, dysphagia, intussusception, obstruction or perforation) or signs of acute or chronic bleeding (23,24).

The diagnosis can be made using endoscopy, in which ultrasonography can be useful in defining the exact localisation and dept of the lesion. Other options are barium radiography, computed tomography (CT) and magnetic resonance imaging (MRI). However, diagnosis might be very difficult and delayed and sometimes requires explorative laparotomy or intraoperative enteroscopy. A biopsy can safely be performed. Histological examination shows a cluster of vessels with an endothelial lining that may appear hyperplastic. According to the size and the type of these vessels, the lesions are devided into 4 groups : capillar, cavernous, mixed capillar-cavernous and mixed haemangioma-lymphangioma. Capillar haemangiomas are usually single lesions, that are weak and may be ulcerated. The vessels often show intraluminal projections. Cavernous hemangiomas are composed of large, thin-walled vessels that may show luminal thrombosis. They can occur in a single or a diffuse fashion. Diffuse cavernous intestinal haemangioma deserves special attention. They occur most commonly in the colon and present with intermittent rectal bleeding, often in childhood. They have a very diffuse growth pattern with possible extension into the genito-urinary tract and retroperitoneum. They may be associated with lymphangiomatosis and protein loosing enteropathy. They are associated with a high morbidity and mortality due to thrombocytopenia that is caused by platelet consumption. Because of the diffuse character of the lesion, surgical intervention can be problematic. Therapeutic angiography has also been tried but is not risk-free (1,4,23). Alpha interferon has shown promising results in a few case reports (4).

Haemangioma' s may also present as part of some syndromes ; i.e. Peutz-Jegher's syndrome, blue rubber bleb nevus syndrome and Klippel-Trenaunay-Weber syndrome.

Peutz-Jegher's syndrome is an autosomal dominant inherited disorder caused by a mutation in a serine/threonine kinase protein gene located on chromosome 19. The syndrome is characterised by mucocutaneous melanin pigmentation (most common on the lips and buccal mucosa, less common on perianal skin and toes) in childhood that can disappear in later life, together with polyposis coli and the presence of haemangioma' s. There is also an increased and cumulative risk of thyroid-, uterine-, ovarian-, testicular-, pancreatic-, breast-, lung- and liver cancer. There also seems to exist an autosomal dominant variant in which there is no polyposis, perhaps due to incomplete penetrance of the mutation or a sporadic mutation in the gene responsible for the syndrome (1,4).

Blue rubber bleb nevus syndrome is a very rare disorder that can be sporadic or inherited in a autosomal dominant pattern. The exact mechanism is not known. Associations have been described with Klippel-Trenaunay's syndrome, Maffucci's syndrome, medulloblastoma, chronic lymphocytic leukemia and renal cell carcinoma (1,4,25).

Patients present with typical skin lesions that have been described as 'blue rubber blebs'. These are bluish, nipple-like structures, covered with a wrinkled skin, that are emptying on pressure and rapidly refilling. 2 other types of skin lesions have also been described : cavernous haemangiomas that may cause compression on or obstruction of vital tissues and blue-black papules or macules that also empty on pressure and may merge with a nevus. Apart from the skin, lesions have also been described in the central nervous system, perineum, eye, liver, spleen, lung, joints, mesenterium and throughout the entire gastrointestinal tract (1,25).

Lesions increase in number from birth onwards. The classical presentation is iron deficiency anaemia in the adult but gastrointestinal bleeding may start at any age from early childhood to middle age (1,4,25). The presence of internal lesions can be confirmed with endoscopy, angiography, CT scan and barium enemas. When biopsied, histological examination shows clusters of dilated capillaries lined with cuboidal cells. Smooth muscle cells and sweat glands may also be present (4,25). Treatment again depends on the mode of presentation and includes therapeutic endoscopy, surgery or pharmacological agents as steroids and interferon alpha (4).

Klippel-Trenaunay-Weber syndrome is a rare, non-hereditary, sporadic disorder affecting children and

young adults. The disease is caused by a vascular abnormality (atresia, hypoplasia or obstruction of a deep venous system, or occasionally an arterio-venous fistula), leading to soft tissue and bony hypertrophy, varicose superficial veins and portwine haemangioma' s, usually sharply demarcated. The lesions are most common on the lower limbs and usually unilateral. They may be complicated by venous leg ulceration, thrombophlebitis, intermittent claudication, lymphedema, gut lymphangiomas, joint dislocation and gait abnormalities. Gastrointestinal involvement is less common. It presents with abnormal serosal vessels and mixed intestinal hemangiomas. Portal vein involvement may lead to portal hypertension. The haemangioma' s may bleed and lead to consumptive coagulopathy. Gastro-intestinal involvement can be confirmed during endoscopy, revealing polypoid vascular projections, especially in the descending colon and rectum. On angiography there may be a slow venous drainage. Multiple phlebolites may be present on radiography. Most common therapeutic modalities are endoscopic sclerotherapy and surgery (1,4).

VII. Angiosarcoma

In contrast to their occurrence in skin, soft tissues, breast, heart , bones, lungs, liver and spleen, angiosarcoma' s are exceedingly rare in the gastrointestinal tract. Most of them are presented in the literature as case reports. They represent about 1,6% of all small intestinal cancers (26-28). Most cases occur after radiotherapy of a gynaecological malignancy, after chemical substitution therapy or as a reaction on foreign material. Angiosarcoma rarely occurs sporadically and also malignant transformation of a haemangioma is exceedingly rare (1,27). The majority of cases are asymptomatic but patients may also present with a mass effect (palpable mass, obstruction and intussusception, perforation or vague pain), anorexia and weight loss and symptoms of gastrointestinal bleeding (26-29). Different diagnostic approaches have been used and include barium enema, CT scan, MRI, ultrasound, Technetium scan, angiography and endoscopy. The diagnosis has to be confirmed by histological examination. Angiosarcoma' s can show a spectrum of well-formed vessels towards haphazard and complex anastomosing slit like channels that are lined by atypical and multilayered endothelium that shows piling up along lumina and may form papillary luminal projections. Other tumours may partly or predominantly be composed of solid sheets of fusiform to epitheloid cells, that usually have large, vesicular nuclei with prominent nucleoli. In these cases, immunohistochemistry might be needed to differentiate the tumour from a carcinoma, melanoma, epitheloid leiomyosarcoma and lymphoma. Angiosarcoma' s can be highlighted with vimentin-, Ulex-, CD31 and CD34 staining. S100- and cytokeratin staining may also be positive. There seems to be an overlap of epitheloid angiosarcoma with epitheloid haemangioendothelioma, perhaps these lesions are both ends of the spectrum of malignant epitheloid vascular neoplasms. Furthermore, the tumors have to be differentiated from primary or metastatic Kaposi sarcoma, basically on morphologic grounds (26,28-30).

Therapeutic strategies are surgical intervention and neoadjuvant chemo-radiotherapy, but the outcome generally is very poor (26,27,29).

VIII. Disorders of connective tissue

Pseudoxanthoma elasticum is an autosomal recessive disorder caused by an alteration on chromosome 16, region p13,1 with a prevalence of 1 in 70000 to 1 in 160000. Occasional cases with an autosomal dominant pattern of heredity have been described. The lesions result from a disturbed elastin metabolism, the exact mechanism of which is yet unclear. There is marked clinical and genetic heterogeneity. Skin lesions are most commonly encountered and described as 'plucked chicken skin'. The retina may show angioid streaks. The mucosa, medium sized blood vessels, heart, kidneys and gastrointestinal tract may also be affected. Vessel involvement results in calcification and failure to contract following injury. This leads to a higher propensity to bleed, especially in the gastrointestinal tract. This bleeding usually originates from the stomach. It may occur in childhood but is more common in the second and third decade. Diagnosis is most often confirmed on skin biopsy. In patients with gastrointestinal involvement, endoscopy may reveal a nodular and friable gastric mucosa with yellow cobblestone-like plaques. Similarly, yellowish plaques may be encountered on colonoscopy. If angiography is performed, often tortuous vessels can be seen. At this moment, treatment of choice for symptomatic cases is angiographic embolisation or surgery (1,4).

Ehlers-Danlos's disease is a group of at least 10 disorders, with marked variation in severity. There are several modes of inheritance, e.g. type IV is autosomal dominant, others are recessive or X-linked. In some types, the specific molecular deficiencies have been elucidated : type IV is caused by a mutation in the collagen 3A1 gene which results in collagen type III deficiency and severe tissue fragility. In type V there is a deficiency in the lysosyl oxidase, in type VI a deficiency in the lysosyl hydroxylase and in type VII a deficiency in the procollagen peptidase. Thus, all types have in common a defect in collagen synthesis leading to hyperelastic and fragile skin and joints with poor scar formation. Furthermore, there is dilatation of blood vessels and valves and dilatations at all levels of the gastrointestinal tract. The disease may be complicated by gastrointestinal and vascular spontaneous rupture. Patients may also present with malabsorption due to bacterial overgrowth and haemorrhages, especially from peptic ulcers, reflux oesophagitis and diverticula. Angiography and surgical

intervention should be avoided. Endoscopic treatment is preferred (1,4).

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