ANGIOMATOSIS

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ABSTRACT

BACKGROUND

Angiomatosis are vascular hamartomas, where there is fair admixture of blood vessels with morphology of veins, arteries and capillaries present in various soft tissues like muscle and deeper plane adipose tissues. Similar lesion is also present sometimes in bone and lower gastrointestinal tract. The angiomatosis may involve both soft tissues and bone, which causes irregular lengthening of the affected limb. Applied aspects are that they may produce thrombosis and gastrointestinal bleeding.

KEYWORDS

Angiomatosis, Soft Tissue and Muscle.

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BACKGROUND

Angiomatosis is an uncommon condition; clinically extensive vascular lesion of soft tissue almost invariably becomes symptomatic during childhood. Because of the nomenclature issues, it has been very difficult to gain a clear understanding of this lesion and in particular to compare the experience derived from one specialty to another. For example, the term angiomatosis has been used in the paediatric literature to refer primarily to multifocal vascular proliferations,[1,2] a criterion that not surprisingly defines a small group of neonates or children with progressive symptoms related to multiple compromise or consumption coagulopathy. Arteriovenous malformation, a term well-founded in the radiologic literature implies functional arteriovenous communications and their sequelae (i.e. hypertrophy of the affected part). An arteriovenous malformation may correspond in some instances to what pathologists term angiomatosis. To promote a more standardised approach to diagnosis, it has been suggested that angiomatosis be used to connote a histologically benign vascular lesion that extensively involves a region of the body or several different tissue types in a contiguous fashion.

The lesions begin during early intrauterine life when limb buds form, grow proportionately with the foetus and consequently affect large areas of trunk or extremity.

The lesions are histologically benign, affect a large segment of the body in a contiguous fashion. Angiomatosis may be of two types:

1. Extensive vertical involvement of multiple tissue planes. (Examples - Subcutis, Muscle, Bone[3])
2. Extensive involvement of tissue of the same type (Example - Multiple muscles)

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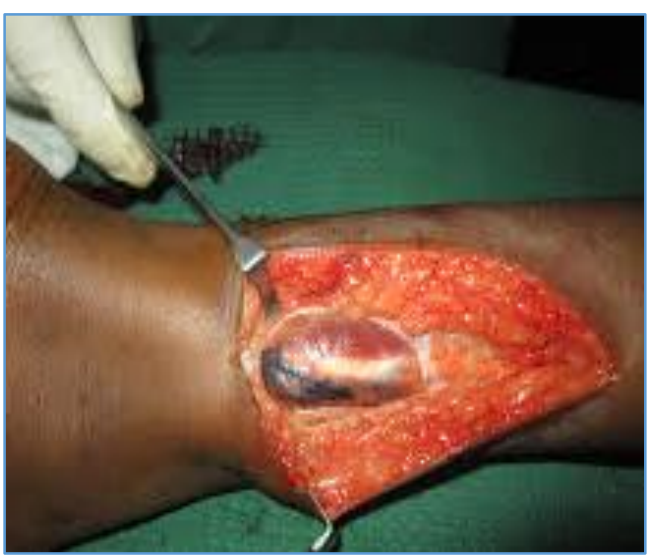
More than half of the patients presents within first two decades of life. Symptoms are diffuse persistent swelling, sometimes associated with pain and discoloration. Hypertrophy, local gigantism and clinical evidence of arteriovenous shunting are rare.

Tumorous lesions often appear as predominantly doughy in nature or as a fatty mass, because of large amounts of interspersed fat.

CASE HISTORY

A 15-year-old female presented to the Surgery OPD with vague mass in left lower one-third of leg, which was clinically diagnosed as haemangioma left leg because of compressibility.

Surgical Picture



Macroscopy

Single grey black, grey brown soft tissue piece measuring 4 x 2 x 1 cm. On cut section, grey black and grey brown haemorrhagic areas identified.

Histology

Collections of large, medium and small-sized blood vessels. Large blood vessels are dilated and filled with RBC's. Some of the vessels have thick walls and the vascular channels are admixed with fibro-fatty tissue. Collection of lymphocytes in between vascular spaces are present, admist the fibrofatty collagenous and skeletal muscle tissue.

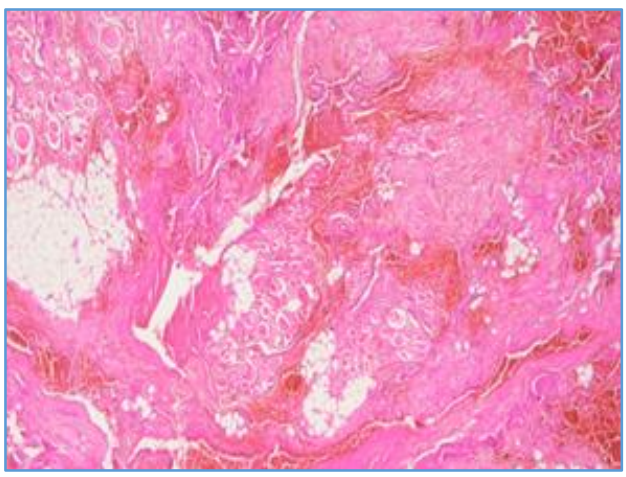


Figure 1. Haematoxylin and Eosin Stained 10x Admixture of Collection of Closely Packed Blood Vessels all of which are Congested

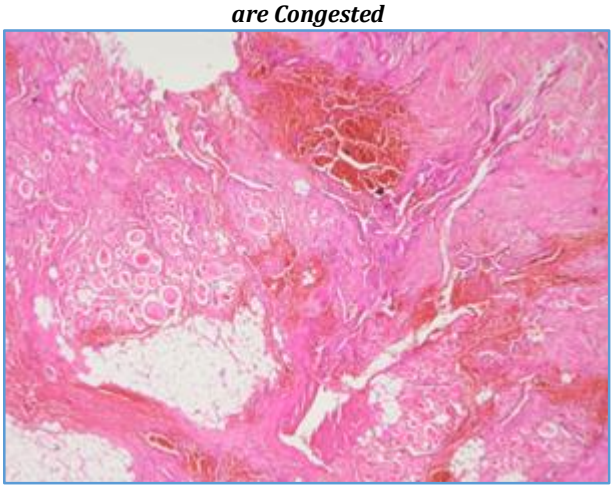


Figure 2. Haematoxylin and Eosin Stained - 40x Another Area showing Dilated and Congested Blood Vessels

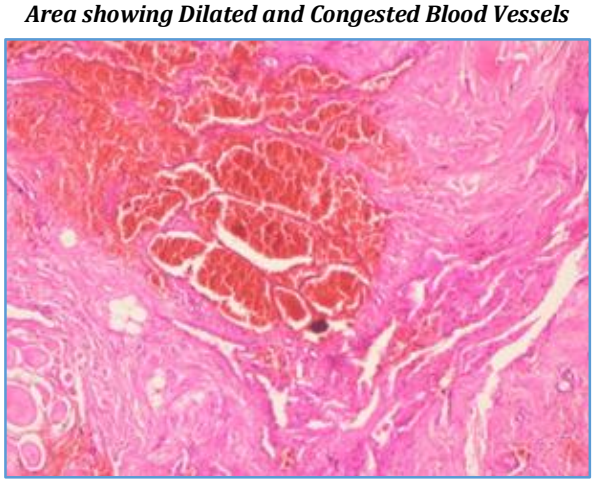


Figure 3. Haematoxylin and Eosin Stained 40x Admixture of Collection of Closely Packed Blood Vessels, all of which are Congested

DISCUSSION

Gross Features

The excised lesions ranged in size from 3 to 26 cm and were usually described as ill-defined with predominantly fatty appearance, marked in areas by red or pink discoloration. Sometimes large-sized vessel can be identified within soft tissue mass, namely fibrocollagenous and muscle tissues; if so, the lesion is described as streaked by fat.

Histologically two different Patterns of Angiomatosis are described[4] namely,

1. The more common pattern seen in most of the cases shows a melange of larger venous, cavernous and capillary-sized vessels scattered haphazardly throughout the soft tissue.

   The venous vessels are remarkable for their irregular, thick walls that have occasional attenuations and herniations. A rather characteristic feature of these veins is the presence of small vessels clustered just adjacent to or in the wall of a larger vein.
2. The next pattern occurs in a small number of cases, which is virtually identical to that of capillary haemangioma, except that the nodules of tumour diffusely infiltrate the surrounding tissue.[5,6]

Prominent amount of fat present in these lesions led previous authors to use the term infiltrating angiolipoma, suggesting that angiomatosis is probably best regarded as a more generalised mesenchymal proliferation. Features of diffuse proliferation of glomus cells in addition to the vessels are also seen.

Angiomatosis has no tendency to progress or evolve histologically with time to a more aggressive lesion. Nonetheless, it is clear that the majority of patients with this condition if followed long enough develop recurrences; 90% of patients experienced recurrences and 40% of patients had more than one recurrence in a 5-year period.[7] A somewhat lower recurrence rate was reported in other study. This behaviour contrasts with the recurrence rate of intramuscular haemangiomas, which is usually less than 50%. Although, there has been speculation that recurrence rates may be higher in young children affected with angiomatosis. It appears not to be true.

There is no evidence that such lesions ever progress to frank malignancy, so the goal of therapy is to treat the lesions as conservatively as possible balancing the need for complete surgical extirpation with the morbidity of the procedure.

The behaviour contrasts with conventional intramuscular haemangiomas, which recur in 20% of cases. Distinction between angiomatosis and intramuscular haemangioma is fundamentally based on clinical rather than pathologic criteria. Irregular venous channels with clustered small vessels in their walls are characteristic of angiomatosis and should certainly in small biopsy specimens prompt a dialogue with the clinician concerning the extent of the lesion. Histological features of this lesion are quite characteristic, but not totally specific for this entity. Unlike cutaneous or intramuscular haemangiomas, in which the most common pattern is that of groups of capillary-sized vessels,[8] angiomatosis is characterised by a mixture of vessels of varying sizes haphazardly involving soft tissues.

Present case, it looks like second pattern where large dilated and congested blood vessels simulating cavernous haemangioma are noticed.

CONCLUSION

Patient came for cosmetic reason, which was clinically diagnosed as haemangioma. During the operative period, it was found that the entire mass seemed to be admixed with blood vessel, adipose tissue within muscle.

Histological pattern exhibited angiomatosis. The case is being presented for the reason that vascular anomalies are of various types, which may have involvement in other anatomical sites and hence the clinical significance. These angiomatosis can coexist with angiomatosis of gastrointestinal tract, bone. In the present patient, fortunately there were no such ailments. She was advised for regular followup.

Prologue

The present patient recovered very well and advised regular followup.

Clinically, there was no evidence of any other vascular abnormality in her.

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