Angiomatosis: a case report

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A 19-year-male was involved in a motor vehicle accident where he sustained a closed acetabular fracture and a left posterior hip dislocation. The preoperative CT abdomen showed an incidental finding of a right 8cm adrenal mass with extensive retroperitoneal and mediastinal lymphadenopathy (Figures 1, 2).

Figures 1 and 2. CT Abdomen arterial phase in coronal plane and transverse plane. Findings: Large adrenal mass which shows renal vessel invasion and IVC displacement anteriorly. Tumour is adherent to right crus and posterior diaphragm. Extensive ipsilateral, retrocaval, retrocaval and contralateral para-aortic lymphadenopathy (not shown).

Following orthopaedic fixation at Southland Hospital, the patient was referred to the endocrine surgeons at Christchurch Hospital for further investigations. Preoperative laboratory findings included normal urine and serum catecholamines, aldosterone, cortisol, αFP, βHCG and sex hormones. The provisional diagnosis was a right adrenal adenocortical carcinoma.

The operation involved endocrine, cardiothoracic and vascular surgeons. A subcostal thoraco-abdominal approach was undertaken with aortocaval and mediastinal lymph node resection, ligation of the renal and adrenal vessels and the tumour and kidney dissected en bloc. The diaphragmatic repair was performed using a dual mesh patch. There was further mediastinal lymph node involvement superior to the carina although this was inaccessible to resection with approach used. The lymphadenopathy was considerably more extensive than what was demonstrated on the preoperative imaging.

Histology from the right adrenal gland, right kidney, diaphragm, right crus and mediastinal lymph nodes gave a provisional diagnosis of a kaposiform
haemangioendothelioma. There were 56 lymph nodes in the abdominal dissection although none were implicated in the disease. The tumour was present on the superior margin of the main specimen encompassing diaphragmatic skeletal muscle fibres. The histological specimen was sent internationally for further opinions and after several months and much controversy, a final diagnosis of angiomatosis was made.

Postoperative recovery was smooth with an uneventful overnight admission to ICU. He was an inpatient for a week and discharged stable. The patient was followed up two weeks later and had recovered well. He was not offered adjuvant therapy and further surgery is not currently considered.

Discussion

Angiomatosis is a rare benign vascular lesion which presents during childhood or adolescence. The lesion will slowly and typically develop in a contiguous fashion in single or multiple tissue types (e.g. subcuticular, muscle or bone). The incidence and prevalence of the condition is unknown. Over 50% of the lesions are usually confined to the lower extremities and 72% of cases present within the first 2 decades of life.\(^1\) Familial cases are rare.\(^2\) The clinical presentation is typically with persistent swelling often associated with pain and skin pigmentation. There are characteristic histological features.

The diagnostic workup involves clinical assessment, imaging and histology. CT is the imaging modality of choice and the lesion appears as a non homogeneous mass which has similar features to a sarcoma except dense areas due to thick-walled tortuous vessels.\(^1\) There is often a lot of fat which can confuse the diagnosis. Histology is the diagnostic gold standard.

Typical histological features can assume two patterns.\(^3\) Most commonly, there is an irregular collage of veins, cavernous vascular spaces and capillaries diffusely positioned. The venous vessels have irregular thick walls and arteries are occasionally identified with a variable incomplete muscle layer (Figures 3, 4). The less common histological presentation is characterised with smaller vessels arranged in nodules around a larger vessel which diffusely infiltrates soft tissue, unlike a capillary haemangioma.
Spontaneous regression of the tumour is rare. Treatment options are limited due to rarity of the condition. Conservative therapy, immune modulating therapies (e.g., interferon, steroids and cyclophosphamide) or surgery are acceptable treatment options and have been suggested in the literature. Ideal follow-up time is not known and in the series by Rao and Weiss,\(^1\) patients were followed up from 1-24 years. They found that nearly 90% of patients had recurrences with 40% having recurrences within 5 years and this varies in the literature from 60–90%.\(^3,4\) The high recurrence rate means that these lesions are difficult to treat.

**Conclusion**

This case led to the incidental finding of a rare vascular tumour. Angiomatosis is predominantly a childhood benign lesion and variable in its presentation. The diagnosis is confirmed on histology. There is no gold standard in treatment and conservative and surgical modalities are both supported in the literature.

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**References:**