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Case Report

Abscess, Epidermoid Cyst or Superficial Angiomyxoma?: A Rare Case of Cutaneous Tumour in the Vulva in an 8-year-old Girl

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Abstract A superficial angiomyxoma is a benign, quite rare, cutaneous tumour. It may be a component of the Carney complex or occur sporadically. Superficial angiomyxoma is rarely described in children, although it is mostly seen in middle-aged men. Herein, a mass is presented that was developed in the vulva after allogeneic bone marrow transplantation and is defined as superficial angiomyxoma in an eight-year-old female patient who was followed up with thalassaemia major.

Key words *Children; Cutaneous tumour; Superficial angiomyxoma; Thalassaemia major; Vulvar mass*

Introduction

Superficial angiomyxoma (SAM), was first described by Allen et al in 1988, is an extremely rare, yet poorly understood, benign soft tissue tumour.[1] The tumour, containing a prominent myxoid matrix and fine blood vessels, involves the dermal and subcutaneous layer. Although there is no distant metastasis, local recurrences have been reported with a frequency of 30-40% in cases which excision is incomplete. SAM, which is frequently seen in the trunk, head, neck and extremities, has rarely been described in the genital area. It may appear as nodule, papule or polypoid form.[2]

An interesting and extremely rare case, whose pathology was reported as superficial angiomyxoma, whose mass was excised from the left labia majus was reported.

Case Presentation

An eight-year-old female patient, who was followed up for thalassemia major and underwent allogeneic bone marrow transplantation (BMT), was consulted by the Paediatric Haematology Department because of a painless, non-itchy and persistent mass on the left labia majus 2 weeks after BMT was performed. The mass of the patient that appeared on the left labia majus was first evaluated as an abscess, and antibiotic treatment was given, but there was no change in the size of the mass. In the physical examination of the patient; on the left labium majus, there was a nodular, approximately 1x1 cm in size, soft and non-fluctuating mass with no redness around it (Figure 1). Ultrasonography (US) was requested for the mass. The operation was planned for the patient after the US result was reported as "a cystic lesion 14x8 mm in size, with a dense content was observed (epidermoid cyst?)". The mass

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was completely excised at the operation. The patient, who did not have any problems in the early postoperative period, was discharged.

Off-white-brown mucosal tissue with dimensions of 1.2x0.8x0.7 cm was sent to pathology. The tumour, rich in thin-walled blood vessels, some of which were curvilinear, had the appearance of a vaguely multinodular dermal mass with myxoid lobules composed of bland spindle and stellate cells. The cells had ovoid to round nuclei and delicate cytoplasmic processes. Stromal neutrophilic infiltration was also observed. Cytological atypia and mitotic activity were absent. Although there was no immunohistochemical staining in tumour cells with CD34, while vascular structures were positive, desmin, SMA, oestrogen receptor (ER), progesterone receptor (PR), S-100 were negative. Mucin accumulation was observed in the myxoid stroma with AB-PAS, histochemically (Figure 2). With these informations, the pathology result of the patient was reported as "superficial angiomyxoma".

The patient, who had no complaints during outpatient follow-up and whose pathology result was superficial angiomyxoma, was re-evaluated in terms of Carney complex (CC). It was learned that no thrombus or vegetation was observed in the echocardiography performed before allogeneic BMT. Physical examination revealed no other lesions or pigmentation on the body, and there was no skin disease in the family history.

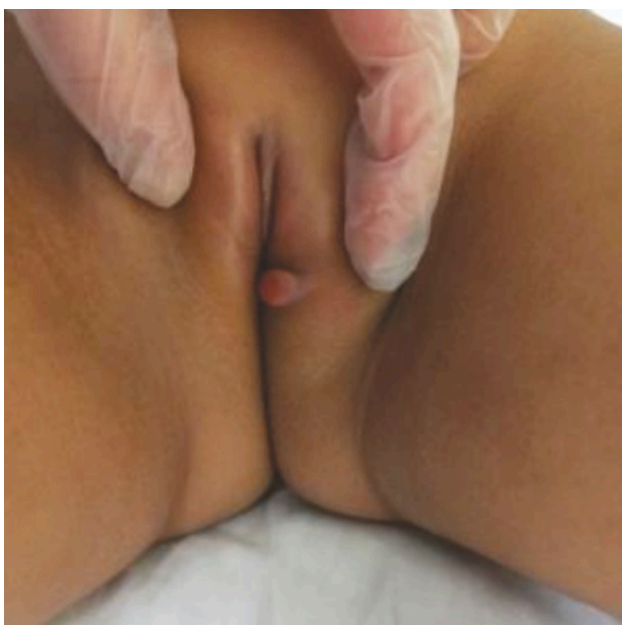

Figure 1 Preooperative examination of patient.

Discussion

Myxomas defined as "true mesenchymal neoplasms consisting of undifferentiated stellate cells placed in myxoid stroma" by Stout in 1948 are called "angiomyxomas" due to the presence of vascular elements.[3] SAM was first described by Allen et al in 1988.[4] Although SAM is frequently seen in men in their 40s, it has also been reported in children and women. It is known that SAM, which is defined in relation to the CC, can also occur sporadically.[5]

CC is an autosomal dominant and rarely, with de novo genetic mutation and a result of mutations in the PRKAR1A gene. CC is a condition characteris=ed by pigmented skin lesions, myxomas (cardiac, skin, breast) and endocrinopathies. Since cardiac myxomas can cause sudden death, every patient with SAM should be examined in terms of CC. Because SAM may appear as the first clinical finding of CC.[6]

SAM is a benign, extremely rare, often painless, slow-growing tumour involving the dermis and subcutaneous tissue. It often occurs in the head, neck, trunk and extremities in the body. In a study by Calonje et al involving 39 patients, it was reported that the dimensions could vary between 1 and 5 cm and the average was 2.3 cm.[7] In the study of Allen et al, it was reported that the majority was between 1-5 cm and could be in sizes ranging from 0.5 to 9 cm.[4] It may occur as papule, nodule, polypoid. In the differential diagnosis, there may be abscess and epidermoid cysts as in our case. In addition, benign or low-grade malignant myxoid lesions such as labial cysts, hernia, Bartholin's or Gardner's duct cysts, aggressive angiomyxoma (AAM), focal cutaneous mucinous, myxoid neurofibroma, myxoid neurofibroma, and myxoid liposarcoma should be considered.[1]

AAM, was first mentioned by Rosai and Steeper in 1983 as a lesion with a high risk of recurrence, which often develops in the pelvic and perineal region in women.[8] AAM and SAM are very similar, however, it is not difficult to make differential diagnosis histologically. AAM is infiltrative and blood vessels are thick-walled. Immunohistochemically, it shows ER and PR positivity. In the histology of SAM, there are spindle cells scattered in the myxoid stroma. Blood vessels are small and thin-walled. Nuclear atypia and hyperchromasia are extremely rare.[2] Stromal inflammatory cells, particularly neutrophils, are common and are not found in other myxoid tumours, so the presence of neutrophils is a clue to a diagnosis of SAM.[7] It may contain epithelial components and it has

been stated that epithelial components are important for recurrence.[5]

In the study of thirty-nine patients by Calonje et al, it was seen that the shortest relapse time was 2 months. It is known that recurrence can be seen with a frequency of 30-40% in cases where surgical excision is not completely performed. Recurrences can also be larger in size than the lesion that first appeared.[7]

The treatment of superficial angiomyxomas is surgical removal of the entire lesion.

Our patient's vulva lesion was initially diagnosed as an abscess due to immune system suppression that occurred during the early stages of allogeneic BMT; however, further testing and treatment were given after the initial course of treatment failed. SAM in thalassaemia major or following BMT has not previously been documented in the

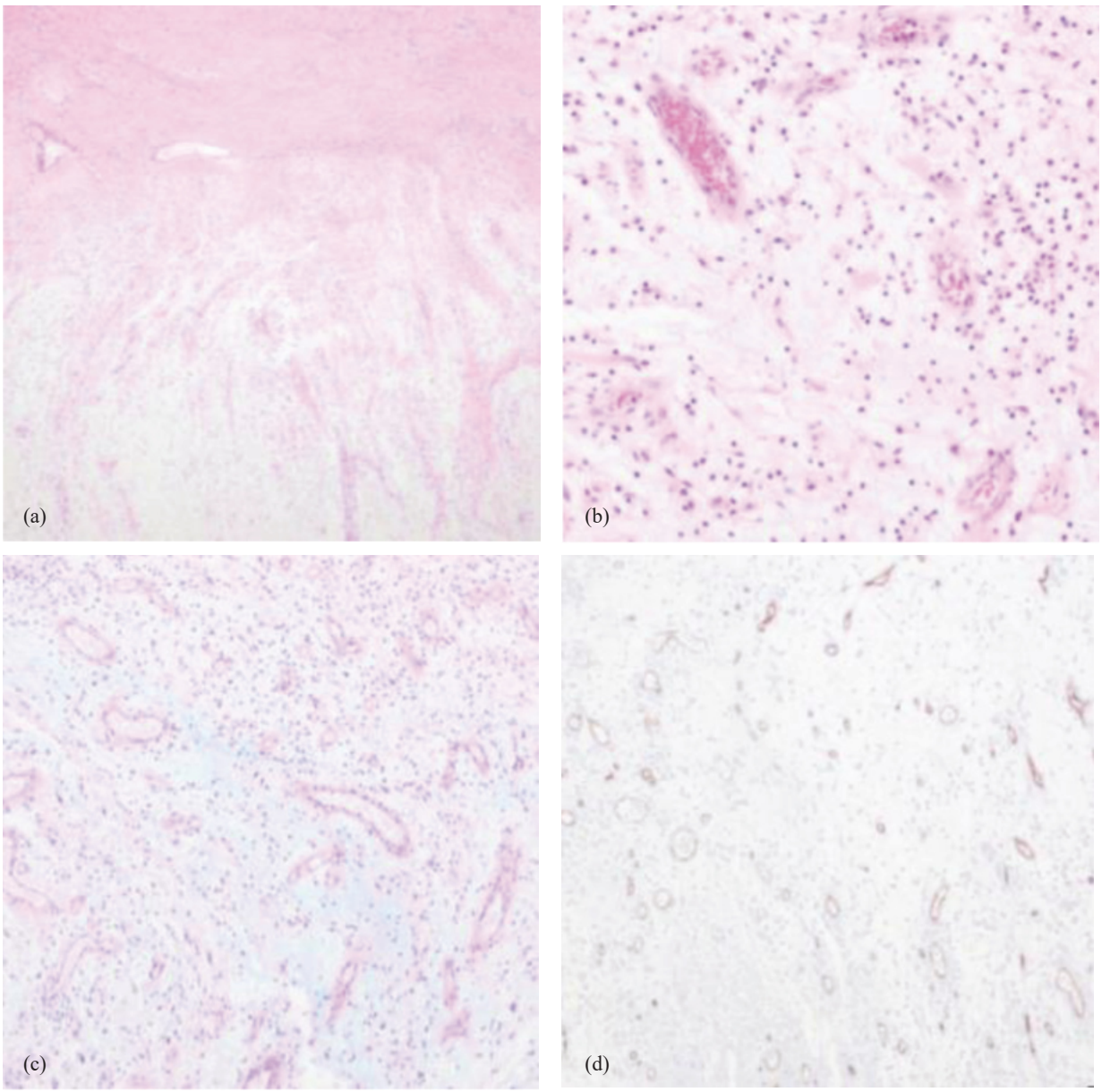

Figure 2 Histopathology of superficial angiomyxoma. (a) Haematoxylin and eosin (H&E stain), x40. Vaguely lobulated lesion with myxoid matrix. (b) Haematoxylin and eosin (H&E stain), x100. Lesion is composed of bland spindle and stellate cells and thin-walled blood vessels set within a copious myxoid matrix. Neutrophil infiltration is also present. (c) Histochemistry, alcian blue-periodic asid schiff (AB-PAS) stained the myxoid stroma. (d) Immunohistochemistry, it is CD34 negative for tumour cells.

literature. In this context, expanding the corpus of knowledge and taking SAM into account when performing a differential diagnosis could be advantageous.

Although SAM is a benign tumour, since it may be a component of CC, it should be carefully examined and the possibility of recurrence should be kept in mind. For this reason, the patient was included in a long-term follow-up program in our clinic.

Declaration of Interest

The author(s) indicated no potential conflicts of interest. No financial or nonfinancial benefits have been received or will be received from any party related directly or indirectly to the subject of this article.

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