Rare disease

Plantar angiomyxolipoma in a child

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Angiomyxolipoma, a lipoma variant with myxoid areas and vascular proliferation was originally described in 1996 and till date has only 12 cases in published literature. Only two cases have been reported in children involving buccal mucosa and knee, respectively. The authors report a case of angiomyxolipoma, on the plantar surface of the left foot, in a 4-year-old male child who presented to our institution in Abha city (Kingdom of Saudi Arabia). The significant differential diagnosis of this neoplasm from other similar lipomatous tumours occurring in adult and paediatric population is discussed. The importance of recognising these tumours lies in their recognition as separate entity and the present case may add to the knowledge, clinical behaviour and prognosis of these less reported lipomatous neoplasms.

BACKGROUND

Lipomas are the most common mesenchymal tumours of adults and are rare in children. By definition, the predominant component is mature adipose tissue but the nomenclature of these neoplasms is vast as per the type of other heterologous mesenchymal elements like fibrolipoma, ossifying lipoma, angiolipoma, myxolipoma, myolipoma or a combination of these as angiolipoma or angiomyxolipoma. The presence of admixture of myxoid component with mature adipose tissue in a background of numerous vascular structures, was first reported as 'angiomyxolipoma' (AML) in spermatic cord.¹ Kim *et al* reviewed 10 cases of AML including their case of AML in thigh of a 9-year-male.² The last reported case of AML was in the oral cavity of a 12-year-old male.³ The clinical outcome was excellent in all cases.

Herein we report the first case of AML on the plantar aspect of left foot corresponding to great toe, in a 4-year-male child, discuss its clinical and histopathological features along with the differential diagnoses from similar adipocytic tumours. The present case may add to the knowledge, clinical behaviour and prognosis of these less reported lipomatous neoplasms highlighting their benign nature and excellent surgical outcome.

CASE PRESENTATION

A 4-year-old boy presented to this hospital and teaching institution located in Abha city of province Asir in Kingdom of Saudi Arabia with a slowly growing painless subcutaneous soft tissue mass located on the plantar surface of the left foot corresponding to the big toe (first metatarso-phalangeal joint), developing over 1-year period. His medical and surgical history was unremarkable and there was no history of trauma at the site of mass. General examination was appropriate for age. Local examination showed a solitary, well circumscribed 2.0×2.5 cm soft tissue mass, which was mobile, not fixed to underlying structures and tender on palpation. The mass was totally excised and a yellow-gray, gelatinous tumour was removed. The

whole specimen was submitted for histopathological examination.

Histopathological findings

Gross: The specimen was composed of a well-circumscribed 2.5×2.0×1.5 cm semisolid tissue mass, surrounded by a thin fibrous capsule.

Cut section: Serial cuts showed yellowish gray surface along with gelatinous areas.

Microscopic examination: H&E stained sections showed a multinodular tumour mass consisting of mature adipose tissue, with myxoid areas having dispersed spindle cells and abundant vascular structures in both lipomatous and myxoid areas (figure 1A–C). The lipomatous component was scattered randomly throughout the tumour and was composed of mature adipocytes without cytological atypia. No lipoblasts were noted. The myxoid component constituted approximately 70% of the whole tumour mass, was diffusely positive with Alcian blue (pH 2.5) and showed few spindle cells with long dendritic cytoplasmic processes (figure 1D). Few scattered histiocytes and mast cells were noted within the myxoid areas. No nuclear atypia or mitotic activity was noted within the myxoid component and MIB-1 proliferative index was less than 1%. The angiomatoid component was noted throughout the myxoid background and appeared predominately as thin walled and to a lesser extent as thick-walled vessels.

Immunohistochemistry

Numerous thin-walled and few thick-walled vessels were demonstrated in the angiomatous component of the tumour by a positive immunoreactivity with CD34 antibody (Dako, Clone QBEnd10). Spindle cells in myxoid area were positive for CD34 (figure 2A). Spindle cells also showed vimentin positivity. The mature adipocytes showed strong immunoreactivity for S-100 (Dako, Z0311) (figure 2B). The above findings led to a conclusive diagnosis of AML.

Figure 1 (A–D) Histopathological features of angiomyxolipoma.

Figure 2 (A, B) Immunopositivity for CD34 and S100.

INVESTIGATIONS

- Haemoglobin: 13.6 gm/dl
- Total leucocyte count: 6600 cells/cu.mm
- Differential Leucocyte count: Polymorphs 67%, lymphocyte 31%, eosinophil 2%.

DIFFERENTIAL DIAGNOSIS

Lipomatous neoplasms commonly involving paediatric population include plantar lipofibroma which is tumour like lipomatous process which involves peripheral nerves and their branches with interspersed fatty tissue among thickened nerve bundles and endoneural as well as perineural fibrosis.⁴ Lipoblastoma and lipoblastomatosis (diffuse lipoblastoma-histologically identical) which almost exclusively involve the paediatric population are another important differential diagnosis. The lesion shows a wide spectrum of cellular differentiation and maturation, encompassing primitive mesenchymal cells, spindle cells, lipoblasts and mature adipocytes, which are arranged

in discrete lobules with myxoid stroma.⁵ However, the majority of mature adipocytes tend to occupy the central part of lobule with lipoblasts arranged peripherally.⁵ Fibrous hamartoma of infancy contains adipose tissue interspersed between fibrous bands but foci of primitive mesenchymal tissue are seen.⁷ Lipofibromatosis occurring in children is recognised by the integral presence of adipose tissue, the predominant tumour component with proliferative fibroblastic elements. Fascicles of fibroblastic tissue traversing the adipose tissue are intimately associated with the septae, preserving the lobular architecture of the adipose.⁹ There is paucity of myxoid elements and vascular proliferation. Lipofibromatosis shows a tendency of infiltration of both the fibrous and lipomatous components into the surrounding structures.⁹

The possibility of entrapment of adjacent adipose tissue by angiomyxoma or by a reactive myxoid lesion is excluded by the random location of scattered adipose tissue throughout the tumour in AML.¹⁰ Myxoid liposarcoma

is rarely superficial in location, has lipoblasts and delicate plexiform capillaries (chicken wire) with no or little scattered positivity for CD34.¹¹ Myxolipoma is characterised by the presence of substantial basophilic mucopolysaccharide material but these myxoid areas show paucity of spindle cell population although prominent vascularisation may be present.^{11 12} Spindle cell lipoma with myxoid stroma has a prominent spindle cell component with scarce vascular component, in addition to the presence of dense ropy collagen fibres.¹³ Angiofibroma is another tumour which may show vascular structures against a oedematous and myxoid background but these vascular structures are surrounded by myofibroblasts.¹⁴

TREATMENT

The tumour was completely excised.

OUTCOME AND FOLLOW-UP

After 6 months of follow up there was no recurrence.

DISCUSSION

Lipomas are the most common mesenchymal tumour of adults¹⁵; but are rare in children.¹⁶ The predominant component in these tumours is mature adipose tissue but proliferation of other mesenchymal elements has led to a vast nomenclature. Different subtypes were described depending on the presence of other components, such as cartilage (chondrolipoma), bone (osteolipoma), muscle (myolipoma), vascular proliferation (angiolipoma) or abundant myxoid changes (myxolipoma).¹¹ The combination of these elements present in the same tumour led to other terms like angiomylipoma and recently AML.

AML, a recent entity was first described by Mai *et al* in 1996 occurring in the spermatic cord.¹ The nine cases reviewed by Kim *et al* documented one case each of AML in spermatic cord, thigh, arm wrist subungual area, gluteal area and hip, along with two cases in scalp.² The age of presentation was from 32 to 69 years.² All the cases were reported in males except for a single case involving the thigh presenting in a 60-year-old female.² Two cases in children were the last reported cases of AML in literature.^{2 3} Kim *et al* reported a case of AML in a 9-year-old boy presenting as solitary painful mass on right medial thigh and described its radiological findings.² The latest addition was AML presenting in the oral cavity of a 12-year-old male.³ The present case occurring in a 4-year-old child, in addition to the last two cases emphasises that although lipomas are rare in children, the angiomylipomatous variant can be encountered in the paediatric age group.

Microscopically, angiolipomas are characterised by variable degree of vascularity as well as variable amount of mature adipose tissue. The term myxolipoma is recommended when substantial basophilic mucopolysaccharide fills the tumour.¹¹ A combination of these three elements in a lipomatous tumour that is, mature adipocytes, paucicellular myxoid areas and abundant vascular proliferation in form of thick and thin-walled blood vessels are described as AML or vascular myxolipoma.^{1 2 11 15} Immunohistochemical studies show that the mature adipocytes are positive for S-100, and the blood vessels

are immunoreactive for CD34 and smooth muscle actin (SMA). The spindle cells in the myxoid areas are immunoreactive for vimentin, and CD34, and non-immunoreactive for S-100, desmin or SMA.^{2 3} The histopathological findings and immunostain positivity were similar in our case.

Learning points

- ▶ AML is a rare lipoma variant which can be found in paediatric population.
- ▶ The tumour is benign in nature and has an excellent prognosis, but requires detailed and meticulous histopathological examination for exact diagnosis.

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Competing interests None.

Patient consent Obtained.

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