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Case Report

Infantile Fibrosarcoma as the Great Mimicker of Infantile Haemangioma on Imaging

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Abstract Infantile fibrosarcoma (IF) is a rare malignant soft tissue tumour in infancy. It can be mistaken as a benign vascular tumour as both share similar clinical presentation and imaging features. We present a 4-month-old girl that initially presented with a small red spot lesion over the right wrist since two weeks of life which had progressively increased in size. The magnetic resonance imaging features were suggestive of a benign vascular tumour that was thought to be an infantile haemangioma. Child underwent total excision of the lesion. However, histopathological examination revealed an infantile fibrosarcoma. We discuss and highlight the radiological features of IF, which often overlaps with vascular benign tumour and also discuss some of the salient features to differentiate between these two diagnoses, as it will lead to different prognosis and management. We believed that in a large soft tissue lesion, whenever the size increases rapidly and disproportionate to the growth of the child, a malignant lesion needs to be excluded and IF should be in the differential diagnosis especially when there is additional presence of intratumoural bleed.

Key words Infantile fibrosarcoma; Infantile haemangioma; MRI

Introduction

Infantile fibrosarcoma (IF) is a rare malignant soft tissue tumour in the paediatric population which has been said to be a great mimicker of infantile haemangioma (IH) either clinically or radiologically.[1,2] About one third of infantile fibrosarcoma present at birth and is often seen in the first 5 years of life.[2] Herein, we present a case report that aims to discuss the clinical and radiological features of IF. We also highlight recent literatures which stress on some differentiating features between IF and IH that can aid in the management of soft tissue lesions in children.

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Methods

A 4-month-old baby girl, who was born at term, presented at two weeks of life with a small red spot over the right wrist, which has rapidly increased in size. On examination, there was a raised erythematous dry skin mass lesion measuring approximately 10 x 8 x 16 cm with minimal serous discharge and contact bleeding (Figure 1). There was no regional or distant lymphadenopathy palpable. Her haemoglobin (Hb) and platelet counts on admission was 10 g/dL and 393x10^6/L respectively. Her coagulation profile, i.e., PT/APTT levels, clotted, but there was no previously documented deranged coagulation profile. She was diagnosed with ulcerated infantile haemangioma in

proliferative phase and was started on syrup propranolol with differential diagnosis of tufted haemangioma.

There was an episode of active bleeding from the lesion. No information was described on the colour change in the case note. Compression was done and subsequently bleeding stopped. Another episode of bleeding had occurred, causing a drop in Hb level to 6.9 g/dL, requiring blood transfusion. Culture sampling from the lesion grew *Staphylococcus aureus*. She was treated with intravenous (IV) cefuroxime for a week. Magnetic resonance imaging (MRI) was performed three months after the initial presentation to further evaluate the lesion. At the time of imaging, the size was unchanged as compared to the size prior to propranolol therapy.

MRI demonstrated a large superficial skin mass centred at the dorsal right wrist. The mass extended superiorly up to the distal third of the radius/ulna level and inferiorly to the proximal carpal bones level. It measured 5.8 cm in maximal dimensions. The mass was isointense compared with muscle on T1 weighted (T1) imaging and hyperintense on T2 weighted (T2) imaging with multiple flow voids within representing intratumoural vessels. On post-contrast, the mass showed homogenous enhancement with prominent vessel at the periphery of the mass, which represented a feeding artery. There was progressive enhancement of the mass on contrast-enhanced dynamic magnetic resonance angiography study, which demonstrated centripetal pattern of enhancement (Figure 2).

The child underwent total excision of the mass lesion with pre-operative diagnosis of infantile haemangioma. The tissue specimen was reviewed by paediatric histopathologist which was reported to show presence of cellular spindle-shaped cells with tumour cells forming irregular fascicles and occasionally arranged in a herringbone pattern. The histopathological examination (HPE) was concluded as infantile fibrosarcoma (Figure 3). Immunohistochemical studies showed the tumour cells were positive to vimentin and negative to Smooth Muscle Actin, desmin, CD34 and S100. However, molecular tests for fusion genes were not performed.

Unfortunately, there was no imaging study to exclude lymph node and distant metastases before the operation since the indication for operation was to stop the bleeding of a lesion that was thought to be benign. Computed tomography (CT) scan thorax was done post-operatively after the HPE came back as IF which showed no distant metastases.

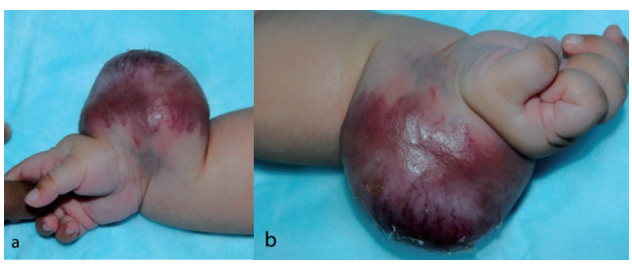

Figure 1 Supination (a) and neutral (b) position of the right hand photos show huge mass at the lateral dorsal right wrist, which has red skin discoloration, central ulceration and prominent vessels at the periphery.

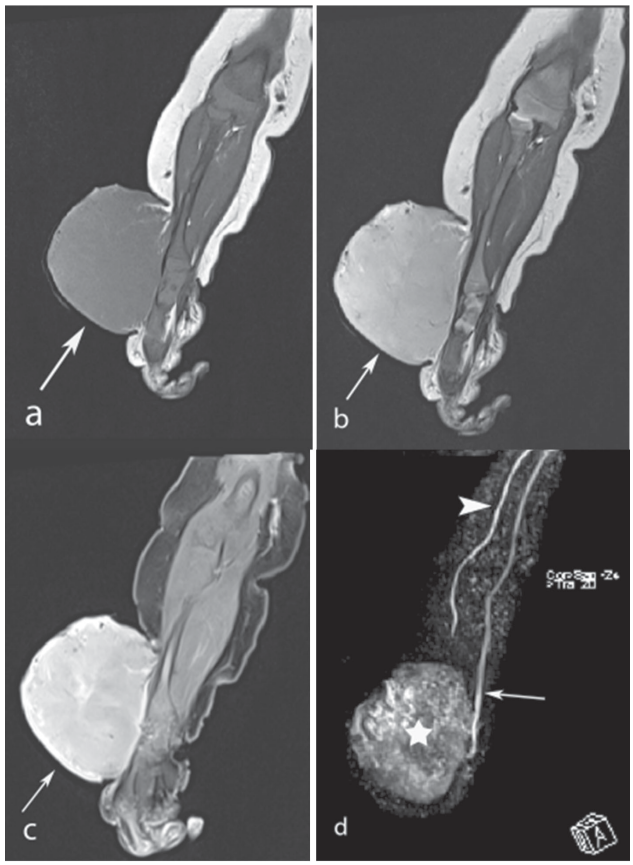

Figure 2 Pre contrast T1WI (a) and T2WI (b), post contrast fat-saturated T1 (c) and maximum intensity projection magnetic resonance angiography (MRA) 3D T1 dynamic contrast enhanced fat-saturated (d) sagittal sections; demonstrating homogenous T1 hypointense and T2 hyperintense mass signal (arrow) and homogenous enhancement post contrast (a-c). (d) The dynamic MRA image shows enhancing mass (star) with an early opacified prominent feeding artery at the periphery (white arrow) likely the ulnar artery and radial artery (white arrowhead), supplying the mass.

Post excision, the patient underwent chemotherapy for a total of 22 weeks (16 cycles IV vincristine and 8 cycles IV actinomycin). She had a repeated MRI of the right forearm and hand 4 months post excision (after 9 cycles of IV vincristine and 5 cycles of IV actinomycin) that favours small residual disease. Another MRI of the right forearm was performed 3 months later after completion of chemotherapy showed no evidence of residual tumour.

Discussions

The majority of soft tissue tumours during infancy, between birth to 12 months of age are pathologically benign, and consist of more than 75%, while 10% and 15% of them are borderline and malignant respectively.[1] Among the malignant mesenchymal tumours in infancy, rhabdomyosarcoma (RMS) accounts for more than 75% of cases as compared to non-RMS, the latter includes undifferentiated sarcomas and infantile fibrosarcoma.[1]

IF is a rare malignant soft tissue tumour in the paediatric population which accounts for 5-10% of all sarcomas in infants less than 1 year of age.[1,2] Infantile fibrosarcoma may present at birth, and until 3 months of age they account for more than 50% of IF cases in a study of 56 patients, also known as congenital fibrosarcoma, or it can develop in the first 5 years of life, but particularly in infants aged less than 2 years.[1-3]

IF usually presents with soft tissue mass on the extremity which accounts for 66% of cases followed by the trunk (25%).[2] The tumour would commonly be large in size at the time of presentation, >5 cm and it shows rapid increase in size.[2,3] The mass sometimes mimics the proliferative phase of haemangioma causing errors in initial clinical diagnosis.[4] The skin changes which includes bluish or reddish discolouration is contributed by intratumoural haemorrhage and necrosis which are also common tumour behaviour that can also be applied to haemangioma.[3,4] However, nothing was described regarding the colour change in the case notes for this patient.

The case showed similar findings to those in the established literature of which the mass appears at 2 weeks of life and rapidly increases in size within a period of three months. There was an episode of ulceration and bleeding causing a significant dropped in haemoglobin level requiring blood transfusion. In view of the presentation, benign vascular tumour such as haemangioma was suspected and the low Hb was thought to be attributed by the intratumoural bleeding. Vascular tumours are known to cause haematological complications such as Kasabach-Merritt syndrome which is a potentially life-threatening coagulopathy, characterised by enlarging haemangioma with severe thrombocytopenia.[3,5] Kasabach-Merritt syndrome (KMS) is unlikely to have been present in this patient as her platelet was normal (no thrombocytopenia) and there was no documented deranged coagulation profile. KMS is commonly seen in proliferating haemangioma; but unfortunately can also be seen in IF.[3,5]

Imaging of IF is usually non-specific.[3,6-8] IF can be confused with benign vascular tumour such as haemangioma on ultrasound and CT.[6] Even though MRI findings are also not specific for IF, it is the modality of choice as it is particularly useful in demonstrating the disruption of soft tissue planes due to its multiplanar capacity, thus helping in pre-operative treatment planning as well as for follow-up.[7] Some of the appearances are similar to infantile or congenital haemangioma (IH/CH) which include heterogenous signal intensity on both T1 and T2 and show variable enhancement on post-gadolinium with the location commonly in the extremities.[3,7,8] A series of imaging features of IF concluded that even though MRI does not show any specific features for IF, this diagnosis need to be suspected especially in case of an infantile soft tissue mass at the extremity that showed intratumoural haemorrhages.[8]

Microscopically, IF has cellular spindle shaped cells with tumour cells most often forming irregular fascicles and occasionally arranged in a herringbone pattern. It is also highly cellular.[4] However, histological diagnosis of IF can carry a diagnostic challenge as it can also mimic

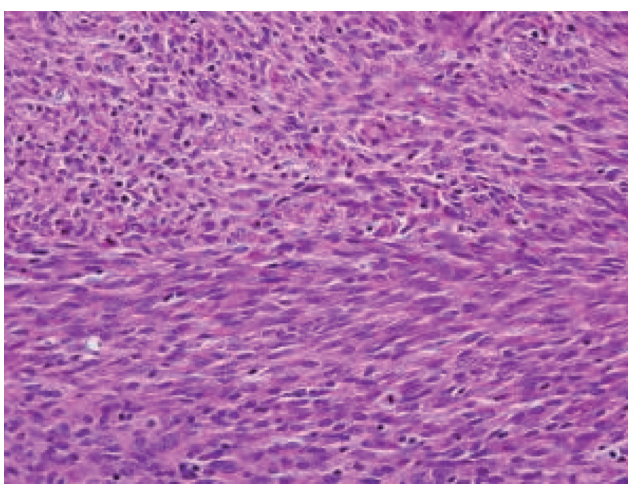

Figure 3 HPE show cellular spindle shaped cells with tumour cells forming irregular fascicles and occasionally arranged in a herringbone pattern consistent with infantile fibrosarcoma.

haemangioma on HPE; which is why molecular studies are helpful in differentiating these two pathologies. Infantile fibrosarcoma demonstrates distinctive reciprocal translocation, t(12;15)(p13;q25), resulting in ETV6/NTRK3 gene fusion, unfortunately this could not be performed in our setting at that point of time.[4]

IF has good prognosis of 80-90% 5 years survival rate as compared to RMS that has poorer prognosis as IF rarely metastasises (8%) despite showing higher risk of local recurrence of up to 50%.[1,2] Therefore, early diagnosis of IF is important to avoid aggressive limb amputation excision and since the prognosis is much more favourable. Definitive surgical resection remains the mainstay of treatment although complete resection is rarely feasible at diagnosis due to the local infiltration and size. First-line chemotherapy such as alkylating agent-free and anthracycline-free regimen has been chosen for inoperable tumours.[2] Chemotherapy toxicity has been manageable with appropriate dose modification.[1] Nevertheless, progression and relapses, mainly local recurrences remain possible despite of the good overall prognosis.[2,9]

Conclusion

A large soft tissue mass in the extremities among infant population has no specific imaging features. Therefore, imaging would not be able to differentiate between a benign vascular mass lesion such as in infantile or congenital haemangioma, or intermediate/malignant mass lesion such as in infantile fibrosarcoma, tufted angioma or kaposiform haemangioendothelioma. Whenever the lesion is rapidly increasing in size and disproportionate to the growth of the child, a malignant lesion needs to be excluded and the risk is increased in the presence of intratumoural bleeding, and IF should be in the differential diagnosis.

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Declaration of Interest

None.

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