J Hong Kong Col Radiol. 2010;13:213-6

CASE REPORT

Inflammatory Myofibroblastic Tumour of the Neck

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ABSTRACT

Inflammatory myofibroblastic tumour is a rare benign neoplasm that has clinical presentations which may mimic malignancy. Most occur in childhood or early adulthood involving the lungs or orbits, but may occur at any age and in any organ system. Preoperative recognition of this quasineoplastic lesion is important, so as to avoid unnecessary radical resection. We report on an infant with an inflammatory myofibroblastic tumour in the soft tissue of the posterior neck, which is a rare site of presentation.

Key Words: *Fibrosarcoma; Granuloma, plasma cell; Inflammation; Tomography, X-ray computed; Ultrasonography*

中文摘要

頸部的炎性肌纖維母細胞性腫瘤

羅彪、方俊仁、仇鴻烈、朱惠邦、麥詠詩、馬嘉輝

炎性肌纖維母細胞性腫瘤是一種罕見的良性腫瘤，其臨床表現與惡性腫瘤很相似。這病主要發生在兒童和年輕人的肺部和眼部，但也可以發生在任何年齡的病人及所有系統上。為避免進行不必要的根治性切除，術前認識類似的腫瘤病灶相當重要。本文報告一名嬰兒的後頸部軟組織炎性肌纖維母細胞性腫瘤，發生的部位很罕見。

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT), popularly known as inflammatory pseudotumour, is a rare benign neoplasm of unknown cause. Most occur in the childhood or early adulthood involving the lungs or orbits, but have been reported at any age and in any organ system. The preoperative recognition of this quasineoplastic lesion is important in order to avoid unnecessary radical resection.[1,2] This IMT occurred in the soft tissue of the posterior neck in an infant, which is a rare location for such a presentation. The clinical, radiological, intraoperative, and histopathological findings are discussed.

CASE REPORT

A nine-month-old baby girl initially presented with upper respiratory tract symptoms and fever (39.2°C), was incidentally found to have a right suboccipital mass of 1 to 2 cm in size. The mass was initially soft, but became firm as it gradually enlarged, but was otherwise asymptomatic and the overlying skin was normal. No discharge or local warmth was found. Over a six-month period, the size of mass reached 5 cm x 4 cm, and there was significant enlargement in the last one to two months. The blood test parameters and inflammatory markers were all normal, except for an elevated erythrocyte sedimentation rate of 24 mm/h. The Epstein-Barr viral titre was normal. No enlarged

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Submitted: 21 Jul 2010; Accepted: 1 Sep 2010.

cervical lymph nodes were palpable.

Fine-needle aspiration cytology (FNAC) of the right suboccipital mass showed spindle cells with scattered small lymphocytes. Computed tomography (CT) performed two days after FNAC demonstrated a large soft tissue mass with slightly lower attenuation compared to adjacent muscle on precontrast images (Figure 1a). On postcontrast images, the mass showed irregular heterogeneous peripheral enhancement with relative hypo-enhancement of its central portion. The mass indented the underlying right upper neck and paraspinal muscles without a distinct fat plane (Figure 1b, c, d). No abnormal calcification or cystic component was found. Tiny bilateral cervical lymph nodes (maximally 0.6 cm in the transverse short axis) were noted.

An ultrasound-guided trucut biopsy was performed a week later. The solid mass appeared well-marginated and positioned within the deep subcutaneous plane closely abutting the right paraspinal muscle (Figure 2a, b). Histology and immunohistological studies yielded a myofibroblastic appearance with an overall clinicopathologic picture favouring IMT (Figure 3)

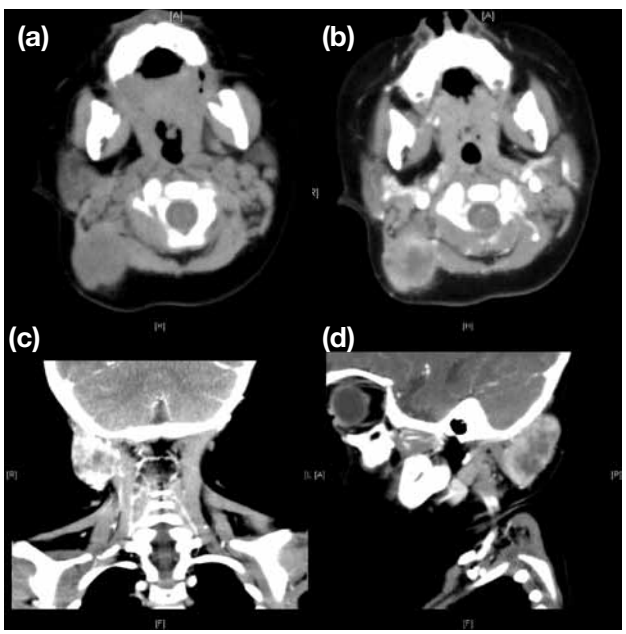

Figure 1. Computed tomography of right suboccipital mass. Axial (a) precontrast and (b) postcontrast images demonstrates a soft tissue mass with heterogeneous enhancement over its periphery, and relative hypoenhancement at its central portion. The deeper musculatures are likely being infiltrated. (c) Coronal and (d) sagittal reformat images of postcontrast scans further illustrate the anatomical relationship.

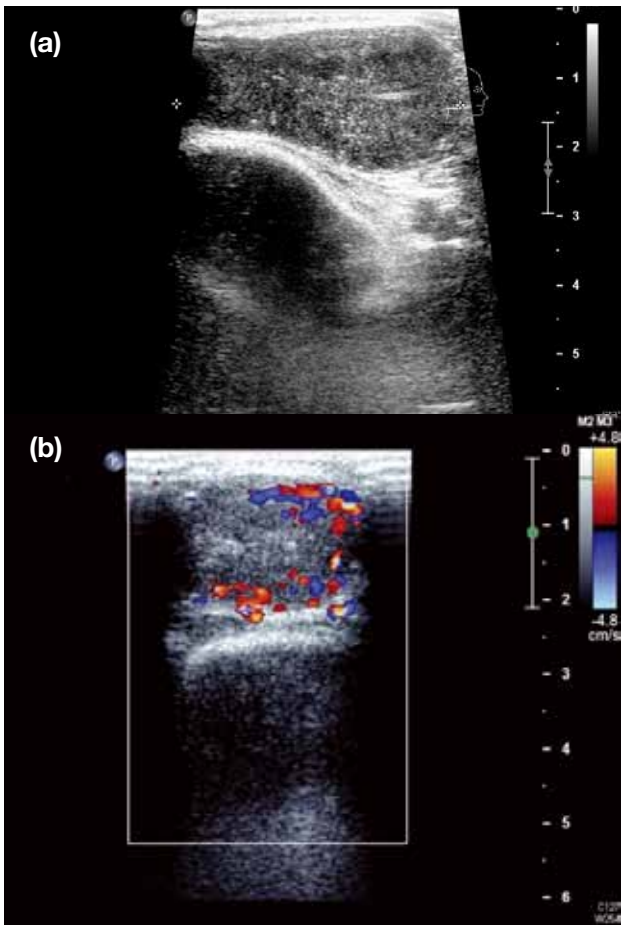

Figure 2. An ultrasound study of right suboccipital mass. (a) Longitudinal scan, the slightly hyperechoic solid mass appears well-marginated and is positioned within the deep subcutaneous plane closely abutting the underlying musculature. (b) Transverse scan with colour Doppler, showing foci of hypervascularity over the peripheral aspect of mass. Trucut core biopsy has subsequently been performed under ultrasound guidance.

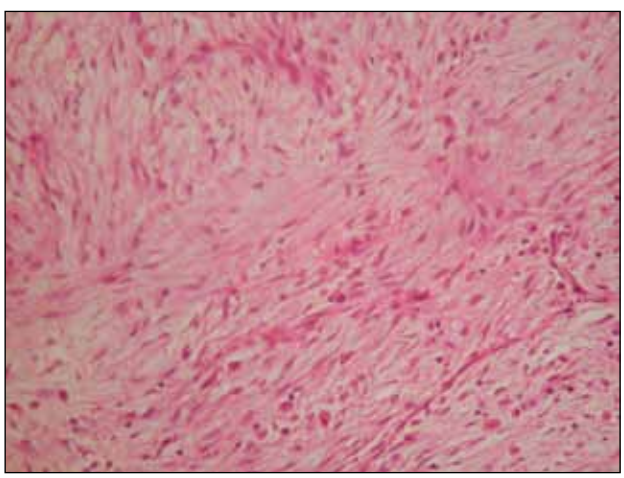

Figure 3. A photomicrograph of the histological specimen: proliferation of spindle cells are seen in a myxoid background, sprinkled with lymphocytes, histiocytes, and plasma cells (H&E, x 200).

as the diagnosis, and nodular fasciitis as a less likely alternative.

One month later, paediatric surgeons excised a hard 4 cm x 3 cm tumour, which was densely adherent to the posterior neck and paraspinal muscles. Its feeder vessels were found and controlled by haemostasis. Complete excision of mass was apparently achieved.

Pathological examinations of the excised specimen confirmed the diagnosis of inflammatory myofibroblastic tumour. Macroscopically, the tumour was an oval tan-coloured solid mass, with no gross area of tumour necrosis. Microscopically, the sections showed low-to-moderate cellular spindle cell proliferation arranged mainly in short fascicular patterns among fibromyxoid or fibrous stroma. This was admixed with a rich inflammatory cells infiltrate, including lymphocytes, plasma cells, histiocytes, foamy histiocytes, and occasional multinucleated cells. The lesion was close to most of the circumferential resection margins. It was non-encapsulated and infiltrated surrounding fatty tissue and skeletal muscle bundles were noted. The deep resection margin was involved. Tumoural necrosis or lymphovascular invasion were not apparent. One benign lymph node was noted. Immunohistological study showed the spindle cells were positive for smooth muscle actin and calponin, and negative for desmin, myogen, Ae1/Ae3, ALK-1, CD34, c-kit, CD21, S100, Melan A and beta-catenin. A small subset of plasma cells were positive for immunoglobulin (Ig) G, while they were largely negative for IgG4, and p53 was weakly positive. The proliferative pool was about 2 to 3% after MIB staining. In-situ hybridisation for Epstein-Barr virus RNA was negative.

The patient had an uneventful postoperative course and to date has remained well during follow-up.

DISCUSSION

IMT is a rare benign neoplasm whose clinical presentation may mimic malignancy. Its aetiology and pathogenesis remain controversial. The term "inflammatory myofibroblastic tumour" was based on electron microscopic and immunohistological findings. This terminology has emerged to distinguish it from the broad category of inflammatory pseudotumours and has now become more common.[1,3,4]

These inflammatory pseudotumours are most commonly found in the lungs and orbits, but have been reported to occur at any age and affect any site in the body. The locations include the maxillary sinus, nasal cavity, tonsils, nasopharynx, larynx, trachea and airways, thyroid, heart, eosophagus, diaphragm, stomach, liver, spleen, pancreas, kidney, adrenal gland, retroperitoneum, mesentery, bladder, testis, central nervous system, fourth ventricle, spinal cord meninges, temporal bone, and the skull base.[2] Mostly, they occur in childhood or early adulthood, and can be asymptomatic or symptomatic (giving rise to fever, weight loss, or a mass effect).[4,5]

The imaging features of IMT vary widely and are usually non-specific. On ultrasonography, the lesions may show variable echogenicity, with ill-defined or well-defined margins. Prominent vascularity may be demonstrated with colour or power Doppler. On contrast CT, the lesions may be homogeneously, heterogeneously, or peripherally enhancing.[1,6-8] On magnetic resonance imaging (MRI), lesions are usually isointense to hypointense on T1-weighted images, relatively hypointense on T2-weighted images, and may show variable enhancement on postcontrast images.[1,2,9] Delayed enhancement may be observed in hepatic and splenic lesions, possibly due to accumulation of extravascular contrast medium in fibrotic areas.[1,6,10]

In extraorbital head and neck locations, their most frequently presenting symptoms are local pain due to obstruction of the involved tract (e.g. pharynx, larynx, or sinuses). Cranial nerve neuropathies have been reported with skull base involvement, as well as perineural or intracranial extension.[7,11] Soft tissues of the neck and cervical lymph nodes are rare sites of IMT occurrence,[1,6] which can be confused with infection, lymphoma, malignant soft tissue tumours, or other proliferative condition such as nodular fasciitis.

CT scan is helpful for defining the location and extent of the lesions. IMT in extraorbital head and neck locations are usually encountered as soft tissue masses with moderate or strong postcontrast enhancement.[2,7] Most masses appear circumscribed, but in some there may be infiltration into surrounding fat.[3,7] On MRI, head and neck IMTs are commonly hypointense on T2-weighted images and show marked homogeneous enhancement on postcontrast images.[9] The low-signal intensity of IMT on T2-weighted images may be a helpful feature to enable differentiation from nodular fasciitis in the head and neck, which is usually hyperintense on T2-weighted images.[12]

The key diagnostic histological findings of IMT include the co-existence of variable numbers of inflammatory cells and spindle cells, consisting of fibroblasts and myofibroblasts with varying degrees of extracellular collagen or fibrosis. This explains such a wide spectrum of synonymous names, including plasma cell granuloma, IMT, inflammatory myofibrohistiocytic proliferation, and inflammatory fibromyxoid tumour.[2,6] Three dominant histological patterns have been described, which can mimic nodular fasciitis, desmoid fibromatosis, or histiocytoma. Histopathological discrimination of these entities may require careful attention to the clinical course, and the additional immunocytochemistry investigations.[3,11]

Corticosteroid therapy is the primary treatment for orbital lesions. Extraorbital head and neck IMT lesions are less responsive to corticosteroids. With exception of several reported cases of spontaneous regression or complete response to medical therapy, for most extraorbital disease the treatment of choice is complete surgical resection.[2] The prognosis is generally favourable if complete resection is achieved; malignant transformation and remote metastasis are rarely encountered.[1,5] Radiotherapy has been tried in those with unresectable disease. Chemotherapy (cyclosporine, methotrexate, azathioprine, and cyclophosphamide) has no significant role.[2]

In conclusion, the clinical presentation of IMT can mimic malignant growths. Although radiological differentiation from malignant lesion is not clear-cut, familiarity with the typical manifestations of IMT is crucial. Before surgery, it is important to consider this entity in the differential diagnosis of soft tissue tumours, as radical resection could be avoided if the diagnosis can be established.[1,2] A histopathological diagnosis (for extraorbital lesions) is valuable for the confirmation of the diagnosis and to exclude malignancy or some other treatable disorder. Early steroid treatment should be instigated in the appropriate clinical context and depending on the tumour's location.[6,7]

ACKNOWLEDGEMENTS

We would like to thank Dr MC To (Resident, Pathology Department, Princess Margaret Hospital) and Dr WS Lam (Associate Consultant, Pathology Department, Princess Margaret Hospital) for providing us the histological photomicrograph for correlation.

REFERENCES

1. Park SB, Lee JH, Weon YC. Imaging findings of head and neck inflammatory pseudotumor. AJR Am J Roentgenol. 2009;193:1180-6.
2. Narla LD, Newman B, Spottswood SS, Narla S, Kolli R. Inflammatory pseudotumor. Radiographics. 2003;23:719-29.
3. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol. 1995;19:859-72.
4. Coffin CM, Dehner LP, Meis-Kindblom JM. Inflammatory myofibroblastic tumor, inflammatory fibrosarcoma, and related lesions: an historical review with differential diagnostic considerations. Semin Diagn Pathol. 1998;15:102-10.
5. Park SB, Cho KS, Kim JK, et al. Inflammatory pseudotumour (myoblastic tumour) of the genitourinary tract. AJR Am J Roentgenol. 2008;191:1255-62.
6. Gunny RS, Akhnar N, Connor SE. CT and MRI appearances of inflammatory pseudotumour of the cervical lymph nodes. Br J Radiol. 2005;78:651-4.
7. De Vuysere S, Hermans R, Sciot R, Crevits I, Marchal G. Extraorbital inflammatory pseudotumor of the head and neck: CT and MR findings in three patients. AJNR Am J Neuroradiol. 1999;20:1133-9.
8. Abehsera M, Vilgrain V, Belghiti J, Fléjou JF, Nahum H. Inflammatory pseudotumor of the liver: radiologic-pathologic correlation. J Comput Assist Tomogr. 1995;19:80-3.
9. Gasparotti R, Zanetti D, Bolzoni A, Gamba P, Morassi ML, Ungari M. Inflammatory myofibroblastic tumor of the temporal bone. AJNR Am J Neuroradiol. 2003;24:2092-6.
10. Nam KJ, Kang HK, Lim JH. Inflammatory pseudotumour of the liver: CT and sonographic findings. AJR Am J Roentgenol. 1996;167:485-7.
11. McKinney AM, Short J, Lucato L, SantaCruz K, McKinney Z, Kim Y. Inflammatory myofibroblastic tumor of the orbit with associated enhancement of the meninges and multiple cranial nerves. AJNR Am J Neuroradiol. 2006;27:2217-20.
12. Kim ST, Kim HJ, Park SW, Baek CH, Byun HS, Kim YM. Nodular fasciitis in the head and neck: CT and MR imaging findings. AJNR Am J Neuroradiol. 2005;26:2617-23.