International Surgery Journal
Pandya B et al. Int Surg J. 2015 May;2(2):270-273
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902
DOI: 10.5455/2349-2902.isj20150529

Case Report

Calcific Inflammatory myofibroblastic tumour in the retroperitoneum of a child: a case report

Bharati Pandya*, Raju Gore, Ravinder Narang

Department of Surgery, MGIMS, Sevagram, Wardha-442102, Maharashtra, India

Received: 05 April 2015
Accepted: 22 April 2015

***Correspondence:**
Dr. Bharati Pandya,
E-mail: bharati_pandya@rediffmail.com

ABSTRACT

Inflammatory Myofibroblastic Tumor (IMT) is a rare benign pseudosarcomatous lesion, characterized by inflammatory infiltrates admixed with myofibroblastic spindle cells. Initially only pulmonary locations were described, now extra-pulmonary sites are also well recognized. We present a case of a 3 year old female child who presented with history of flank pain, abdominal mass and intermittent fever of 1 year duration. Provisional diagnosis was neuroblastoma, but it turned out to be a retroperitoneal IMT with calcification. The patient was managed with surgical resection of the tumor and post-operative treatment with steroids as part of the tumour adherent to the inferior vena cava could not be resected. IMT is an uncommon neoplasm of uncertain biological potential. Both, retroperitoneal location and calcifications are rare presentations. Complete surgical resection remains the mainstay of treatment. Possible treatment of residual and recurrent tumours could be with steroids and anti-inflammatory drugs. Selected cases may need chemotherapy, the efficacy of which is not well documented. It has close resemblance to calcific fibrous pseudo-tumour and we have presented the differentiating features from review of literature.

Keywords: Inflammatory myofibroblastic tumour, Retroperitoneum, Immunohistochemistry, Calcific fibrous pseudo-tumour

INTRODUCTION

Inflammatory Myofibroblastic Tumour (IMT) defines a histologically distinctive lesion with uncertain behavior and denotes a pseudosarcomatous inflammatory lesion that contains spindle cells, myofibroblasts, plasma cells, lymphocytes and histiocytes.[1] Its biological behavior varies from frequently benign lesions to more aggressive variants. IMT occurs commonly in the lung. Extra-pulmonary IMT in children have been described in the mesentery, omentum, liver, bladder, mediastinum, head and neck, extremities, appendix, and kidneys, with the largest tumours occuring in the abdomen and retroperitoneum.[2] Retroperitoneum is a rare site. Calcifications too are rare and confuse this tumor with another distinctive entity calcific fibrous pseudotumour. CFPs have some distinct characteristics which differentiate them from IMTs, though some believe them to be a late sclerozing stage of IMT.[3] IMT frequently recurs and rarely metastasizes[1]. Best proven modality of treatment is surgery.[1] Anti-inflammatory drugs in children have been suggested and chemotherapy for indolent cases has been tried but without documented success.[4,5]

CASE REPORT

A 3 year old female child presented to us with abdominal pain, intermittent fever and gradually progressive abdominal mass of 1 year duration. She was taking

treatment with her local practitioner for urinary tract infection. On examination a hard mass was palpated in the right lumbar region which was fixed to the psoas muscle. Plain X-ray of abdomen showed a calcific round mass in the right lumbar region. Ultrsonography and CT scan revealed a right sided retroperitoneal mass of 8.5 cm x 6.5 cm x 5 cm. with calcifications. Kidney was displaced supero-laterally. Tumour was abutting the inferior vena cava and was buried in the psoas muscle. Aspiration cytology was inconclusive. Surgery was performed. Part of the tumour adherent to the inferior vena cava infero-medially could not be resected. Histopathology reported, inflammatory myofibroblastic tumor of retroperitoneum with calcification. Tumor cells were positive for desmin, vimentin and actin. Post operatively patient was put on steroid therapy and ultrasound done 3 months later showed no evidence of tumour. Follow up is planned with CT scan for recurrence and subsequent tapering of steroids.

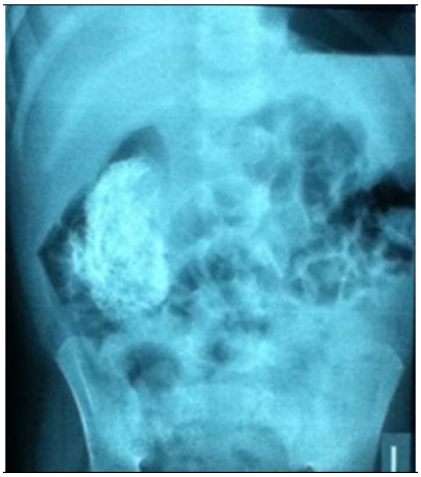

Figure 1: Depicting calcified mass lesion on X-ray which looks like the kidney on plain X-ray.

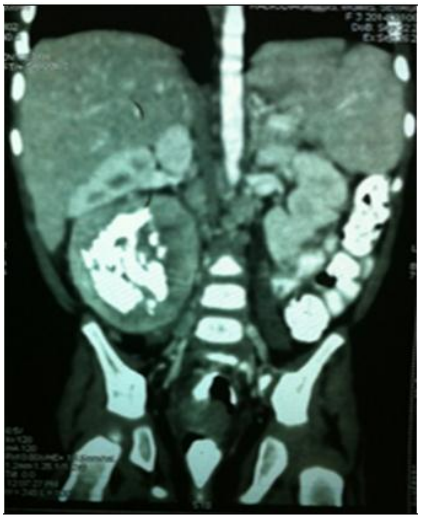

Figure 2: CT scan shows the supero-lateral displacement of the lower pole of kidney and the calcified mass below it on the right side.

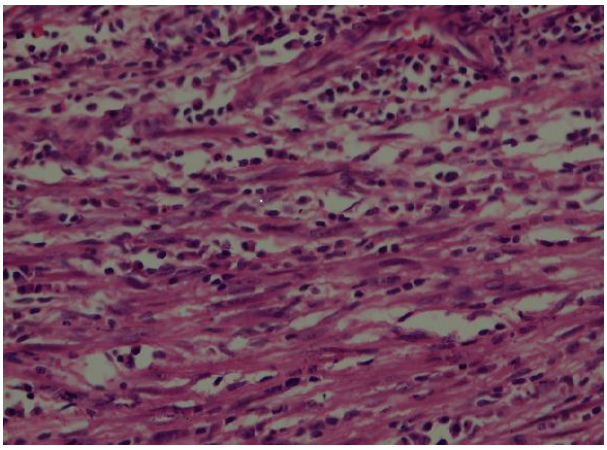

Figure 3: Simple H & E staining of the calcific IMT, Immuno-histochemical staining showed strong positivity for smooth muscle actin (immuno-peroxidase staining).

DISCUSSION

Inflammatory myofibroblastic tumour was first described by Brunn in 1939.[6] The histopathological definition of an IMT is a distinctive neoplasm composed of myofibroblastic mesenchymal spindle cells accompanied by an inflammatory infiltrate of plasma cells.[7] Tissue samples obtained by fine-needle or tru-cut biopsies, and even analysis of perioperative biopsies, are not enough to establish a diagnosis. The pathologist usually asks for the whole specimen.[1,8] In our case, FNAC was inconclusive. Our final diagnosis was only possible after the evaluation of the whole mass and the detection of neoplastic growth of myofibroblastic spindle cells on an inflammatory background with dystrophic calcifications. Currently, surgery is the mainstay of treatment for IMTs.[1,8] Complete removal of the tumour generally provides resolution of all symptoms. However, tumours in intra or retroperitoneal locations tend to invade adjacent structures, preventing curative resections and breeding local recurrences.[8] In our case, visible residual tissue was left behind. Unfortunately, chemotherapy and radiotherapy are not successful in most patients.[2, 9.] Recently, researchers have published promising results with anti-inflammatory agents and anti-tumour necrosis factor-α binding antibodies.[2,9]. Additionally, Berger and colleagues[9] reported the first case of a bladder inflammatory myofibroblastic tumour that responded to an anti-inflammatory regimen (prednisone and Cox-2 inhibitor) even before surgical extirpation.

Calcifying Fibrous Pseudotumor (CFP) is another distinct entity characterized by a predominantly lymphoplasmacytic infiltrate with abundant hyalinized collagen and psammomatous or dystrophic calcifications. These lesions were reported initially as "childhood fibrous pseudotumor with psammoma bodies".[10] The term 'calcifying fibrous pseudotumor' was coined by Fetsch et al. in 1993.[4] The cause and pathogenesis are unclear, but it has been postulated that CFP may

represent a sclerosing end stage of Inflammatory Myofibroblastic Tumor (IMT).[3] Simple excision with a margin of normal tissue is the treatment of choice. The risk for local recurrence is low.[3]

Both IMT and CFP were initially considered to be lesions of young children. Fetsch et al.[4] described the occurrence of CFP in patients between 1 and 33 years of age (mean age, 16.2 years), with a slight predilection for females. CFP usually presents as a mass in an otherwise healthy patient; whereas IMT patients may have other symptoms or signs such as fever, pain, weight loss, malaise, anemia, thrombocytosis, increased sedimentation rate, and hypergammaglobulinemia.[11] Distinction from IMT may be difficult because; spindle cells, an inflammatory infiltrate, and sometimes calcifications characterize both CFP and IMT. Coffin et al.[7, 11] documented IMTs associated with local recurrence. In contrast, CFP has an excellent prognosis, with recurrences being rare.[10] The spindle cells of IMT react intensely against antibodies for muscle-specific actin and desmin, which points to the myofibroblastic nature of the cells. Kalisha et al.[10] in their study have reported that none of the CFPs stained with muscle-specific actin, smooth muscle actin, or desmin.For clarifications, we reproduce the tabular comparision between these two entities studied extensively by Kalisha et al.[10]

Table 1: Showing comparison between inflammatory myofibroblastic tumor and calcifying fibrous pseudotumor (as reported by Kalisha et al.[10]).

Characteristics	IMT	CFP
Age of patients	Children	Young adults
Sex M:F ratio	1:1.3	1:1
Predominant sites	Lungs, abdomen and retro-peritoneum	Soft tissues
Duration of symptoms	Short	Long
Systemic symptoms	Present	Absent
Laboratory abnormalities	Sometimes	no
Recurrences	Occur	Rare
Metastasis	Rare	Never
Histology: Architecture	Multi-patterned	Uniform
:Calcifications	Ocassional	Always
Immunohistochemistry		
:Factor VIII	Intense	Rare/focal
:Factor XIII	Rare	Positive
:CD34	Variable	Negative
:ALK	Common	Rare/focal
:Smooth muscle actin	Strong and diffuse	Negative
:Muscle specific actin	Strongly positive	Negative
:Desmin	Positive	Rare/focal
:Chromosomal patterns	Yes, 2p22-24	unknown

In our patient the main confusion was with CFP. The clinical symptoms of mass with fever, earlier presumed to be urinary infection was a clue which was not taken pre-operatively, which abated post-operatively. Histopathology confirmed calcific IMT with positive staining for muscle specific actin, smooth muscle actin and desmin. The residual tumour in view of retroperitoneal location and histopathology of IMT was treated with anti-inflammatory treatment in the form of steroids, to avoid local recurrence of the tumour.

CONCLUSION

IMTs are rare tumours with distinct histopathology. Retroperitoneal location of this tumor is also rare. Rarer still are calcifications in IMT. In the reported case, the presence of calcification led to the confusion of a neuroblastoma or CFP, pre-operatively. Final histopathology confirmed an IMT with calcification. The CFPs have features including their biological behavior and prognosis distinct from IMTs. Some CFPs may have similar features with IMTs, and it is important to separate those cases in children, especially those arising from the soft tissue and those occurring in the retroperitoneum. Tissue biopsy is essential along with immunohistochemical studies to confirm the diagnosis. IMTs frequently recur, but rarely metastasize. In residual tumors further follow up could be with anti-inflammatory drugs to preventive inoperable recurrences.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Milos Bjelovic, Marjan Micev, Bratislav Spica, Tamara Babic, Dragan Gunjic, Aleksandra Djuric, et al. Primary inflammatory myofibroblastic tumor of the stomach in an adult woman: a case report and review of the literature. World J Surg Oncol. 2013;11:35.
2. Hojo H, Newton WA Jr, Hamoudi AB, Qualman SJ, Wakasa H, Suzuki S, et al. Pseudosarcomatous myofibroblastic tumor of the urinary bladder in children: a study of 11 cases with review of the literature. An Intergroup Rhabdomyosarcoma study. Am J Surg Pathol. 1995;19:1224-36.
3. Van Dorpe J, Ectors N, Geboes K, D'Hoore A, Sciot R. Is calcifying fibrous pseudotumor a late sclerosing stage of inflammatory myofibroblastic tumor? Am J Surg Pathol. 1999;23:329-35.
4. Fetsch JF, Montgomery EA, Meis JM. Calcifying fibrous pseudotumor. Am J Surg Pathol. 1993;17:502-8.
5. Germanidis G, Xanthakis I, Tsitouridis I, Zaramboukas T, Kiskinis D, Konstantaras C, et al. Regression of inflammatory myofibroblastic tumor of the gastrointestinal tract under infliximab treatment. Dig Dis Sci. 2005;50:262-5.

6. Brunn H. Two interesting benign lung tumors of contradictory histopathology. J Thorac Surg. 1939;9:119-31.
7. Coffin CM, Hornick J, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol. 2007;31:509-20.
8. Ozgur Firat, Safak Ozturk, Taner Akalin, Ahmet Coker. Inflammatory myofibroblastic tumour. Can J Surg. 2009 Jun;52(3):E60-1.
9. Berger A, Kim C, Hagstrom N, Ferrer F. Successful preoperative treatment of pediatric bladder inflammatory myofibroblastic tumor with anti-inflammatory therapy. Urology. 2007;70:372.e.13-5.
10. Kalisha A. Hill, Frank Gonzalez-Crussi, Pauline M Chou. Calcifying fibrous pseudotumor versus inflammatory myofibroblastic tumor: a histological and immunohistochemical comparison. Mod Pathol. 2001;14(8):784-90.
11. Coffin Cheryl M, Watterson Jan BA, Priest John R, Dehner Louis P. Extrapulmonary Inflammatory Myofibroblastic tumor (inflammatory pseudotumor) a clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol. 1995 Aug;19(8):859-72.

DOI: 10.5455/2349-2902.isj20150529
Cite this article as: Pandya B, Gore R, Narang R. Calcific Inflammatory myofibroblastic tumour in the retroperitoneum of a child: a case report. Int Surg J 2015;2:270-3.