CASE REPORT

Intra-abdominal inflammatory myofibroblastic tumour (IMT) mimicking gonadal mass in a male infant with undescended testes

Sanjeev Kumar Shukla, Shivya Parashar

Radiodiagnosis, Government Medical College, Kota, India

Correspondence to

Dr Shivya Parashar,
shivya.parashar205@gmail.com

Accepted 27 April 2017

SUMMARY

Inflammatory myofibroblastic tumour (IMT) is a rare benign neoplastic tumour, originally described as an inflammatory pseudotumor. Only a single case of IMT in the spermatic cord associated with undescended testes has been reported. We present a rare case of abdominal IMT in a male infant with undescended bilateral testes. The abdominopelvic mass was suspected to be a gonadal malignancy due to empty scrotal sac but proved to be otherwise on histopathology. Mass was completely resected and orchiopexy was performed for both undescended normal-appearing testes. The patient developed a recurrent mass 2 months later involving the left spermatic cord and the left testicle. Radical surgery was performed with removal of the left testicle and spermatic cord. The patient was disease-free at 1-year follow-up.

BACKGROUND

Only one case of an inflammatory myofibroblastic tumour (IMT) of spermatic cord with undescended testes has been reported, none in infant. Radiological identification of this tumour is important to decide a management guideline. We present this case to share radiological appearances of a primary abdominopelvic IMT in an infant with undescended testes and recurrent genital IMT following surgery.

CASE PRESENTATION

A 3-month-old male patient presented to the department of radiology with rapid progressive abdominal swelling for 2 months, beginning with a nodular swelling in the left inguinal region.

INVESTIGATIONS

Laboratory tests showed elevated neutrophil count ($6.1 \times 10^9/L$) and C reactive protein (16.4 mg/L). Ultrasonography (USG) of the abdomen-pelvis revealed a large heteroechoic mass in the abdominopelvic cavity with remarkable internal vascularity; part of the mass was herniating into the left inguinal canal. The origin of the mass could not be identified on USG imaging. Both scrotal sacs were empty, and bilateral testes were not identified in the inguinal canal region or pelvic cavity.

A contrast-enhanced CT (CECT) of abdomen-pelvis was performed. Non-contrast CT revealed a large (10×10 cm) homogeneous hypodense, lobulated mass in the abdominopelvic

cavity. The mass showed strong heterogeneous enhancement on postcontrast imaging (figure 1). Posterior displacement of the left kidney, inferior mesenteric vein and posterolateral displacement of descending colon suggested the intraperitoneal location of the mass. Multiple enhancing vessels were seen within the mass lesion. A lobe of the mass was herniating into the left inguinal canal (figure 2). Bilateral testicles were not present within the scrotal sac and inguinal canal (figure 3). The testes could not be identified in the abdominopelvic cavity and other ectopic locations. No metastatic deposits were found in the abdominopelvic cavity and vertebral column. Chest X-ray of the patient was normal. On the basis of imaging appearances of the mass and non-visualisation of both the testicles, differential diagnoses of germ cell tumour and intraperitoneal mesenchymal tumour were made. Fine needle aspiration cytology from the mass revealed spindle cell tumour.

Laparotomy was performed on the next day. A lobulated intraperitoneal mass was found in the abdominopelvic cavity, partly herniating into the left inguinal canal as described on radiological imaging. The mass was resected and removed completely (figure 4). Both testes were identified in the abdominal cavity, separately from the mass and appeared normal. Both testes were replaced back into the scrotal sacs and an orchiopexy was performed. Postoperative USG did not reveal any residual mass in the abdominopelvic cavity. Resected mass was sent for histopathological examination. Histopathology and subsequent immunohistochemistry confirmed this spindle cell lesion as an IMT.

Two months after the surgery, the patient was brought back to the department with a large swelling in the left inguinoscrotal region. Repeat USG and CECT of the patient was performed. USG revealed an ill-defined, heterogeneous, hypoechoic, highly vascular lesion infiltrating the left spermatic cord and the left testis, which was enlarged (figures 5 and 6). CECT image showed a heterogeneously enhancing left inguinoscrotal mass involving the left spermatic cord and the left testis, extending to reach the deep inguinal ring (figure 7). Few enhancing lymph nodes were seen around the mass lesion. No intra-abdominal mass was seen on imaging. Right spermatic cord and right testis were normal. The patient underwent radical surgery

CrossMark

To cite: Shukla SK, Parashar S. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-219745

Figure 1 A 3-month-old male child with an inflammatory myofibroblastic tumour. Non-contrast CT and contrast-enhanced CT abdomen-pelvis coronal reconstructed images show a large multilobulated abdominopelvic mass showing marked heterogeneous enhancement. Part of the mass is herniating into the left inguinal canal (red arrow). Both scrotal sacs are empty (asterisk). Urinary bladder (yellow arrow).

with removal of left testis and left spermatic cord. Diagnosis of a recurrent IMT was confirmed on histopathology.

DIFFERENTIAL DIAGNOSIS

1. Germ cell tumour
2. Rhabdomyosarcoma

TREATMENT

Surgical resection of the mass with bilateral orchiopexy was performed initially. Radical surgery with removal of the left testicle and spermatic cord was performed to remove a recurrent scrotal mass.

OUTCOME AND FOLLOW-UP

The patient developed recurrent mass in the left inguino-scrotal region 2 months after initial surgery. Radical surgery was performed with removal of left testis and left spermatic cord at subsequent visit with a recurrent mass lesion. The patient was disease-free at 1-year follow-up.

DISCUSSION

In 1954, Umiker and Iverson¹ derived the term 'inflammatory pseudotumor' because of clinical and radiological similarity of

Figure 2 A 3-month-old male child with an inflammatory myofibroblastic tumour. Contrast-enhanced CT abdomen-pelvis axial image shows the two lobes of the mass mimicking bilateral gonadal mass in an infant due to empty scrotal sacs.

Figure 3 Contrast-enhanced CT axial image at the level of scrotal sac shows empty scrotal sacs (red arrow).

the tumour with malignant tumours. Because of the variable appearances of IMTs on histopathology, several different terms have been used to describe this tumour, for example, plasma cell granuloma, inflammatory pseudotumor (IPT), inflammatory myofibrohistiocytic proliferation, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, fibrous xanthoma, plasma cell histiocytoma in the lung and inflammatory fibrosarcoma in the urinary bladder.² IMTs are mass-forming neoplastic lesions and can develop in almost every organ such as lung, orbit, liver, pancreas, spleen, in the abdominopelvic region, genitourinary system and retroperitoneum, with lungs and orbits being the two most commonly affected sites.³⁻⁵ Involvement of the genitourinary tract is rare and most commonly seen in the urinary bladder.⁴ Low incidence of genitourinary IMT makes the diagnosis challenging.

IMTs have been reported in patients of all age groups, but children and young adults are more commonly affected, with a slight male predominance.⁶ The cause of IPT is still poorly understood. Previously it was thought to be an inflammatory response to micro-organisms such as *Escherichia coli*, Epstein-Barr virus and Cytomegalovirus, or an autoimmune reaction.⁷ However, many recent studies have demonstrated chromosomal abnormalities of 2p23 and rearrangement of the anaplastic lymphoma kinase (ALK) gene locus in IMT, which favours a neoplastic rather than a reactive inflammatory process for IMT development.^{8,9}

Figure 4 A 3-month-old male child with an inflammatory myofibroblastic tumour. Postoperative image shows a multilobulated solid mass with a smooth outline; the mass was entirely removed.

Figure 5 Follow-up (2 months later) ultrasonography reveals an ill-defined, heterogeneous, hypoechoic, highly vascular recurrent lesion infiltrating the left spermatic cord (asterisk) and the left testis, which is enlarged (red arrow).

Clinical presentation varies widely depending on the specific organ involved. These tumours usually remain asymptomatic in early stages or can present with non-specific constitutional symptoms. IMTs can also present with visible or palpable mass or with symptoms of mass effect due to the local aggressive nature of the tumour, as was in our case. Intestinal bleeding or acute abdomen may follow the rupture or haemorrhage within an abdominal mass.⁸

Various imaging appearances of IMTs have been described at different anatomical locations, and this rare entity could be easily mistaken for a malignant neoplasm anywhere in the body, as in our case mistaken as an intra-abdominal gonadal neoplasm. USG is used as the primary imaging modality to evaluate scrotal masses in an infant. CT and MRI are also used to identify the tumour extension into the abdominopelvic cavity and to localise testes in an infant with undescended testes. USG and CT appearances of IMTs are non-specific and do not allow to clearly differentiate IMTs in the genitourinary system from other common malignancies such as rhabdomyosarcoma or primary gonadal malignancies, especially when the scrotal sacs are empty, as was in this case. On ultrasound, IMT can appear hypoechoic or hyperechoic mass with ill-defined or well-defined borders, and often shows increased vascularity on colour or power Doppler.^{9 10} CT also has variable appearances; lesions can be isodense or hypodense on non-contrast imaging compared with the muscles.^{9 10} On MRIs, IMTs usually present with hypointense lesions on both T1-weighted and T2-weighted images, due to the fibrotic nature of IMTs.¹⁰ On CECT and MRI, IMTs show a homogeneous, heterogeneous or peripheral with late prominent central enhancement. Prominent delayed enhancement is seen due to the presence of fibrosis within the lesion. Hypointensity on T2-weighted images could be used as a distinguishing feature

Figure 6 Colour Doppler ultrasonography reveals the high vascular nature of the lesion.

Figure 7 Follow-up (2 months later) contrast-enhanced CT abdomen-pelvis coronal reconstructed image shows a large, heterogeneously enhancing recurrent left inguinoscrotal mass involving the left spermatic cord and the left testis, extending to reach the deep inguinal ring. Few enhancing lymph nodes are present around the mass lesion. Right testis and spermatic cord are normal (red arrow).

from malignant neoplasms, which are usually hyperintense on T2-weighted images.

Definitive diagnosis of IPT is usually made by histopathology and immunohistochemical analysis after surgical removal of the mass. Three different patterns of IMTs have been described on histopathological examination, most common being myofibroblastic spindle cells with inflammatory infiltrates predominantly containing lymphocytes, histiocytes, plasma cells and eosinophils.⁸

Surgical resection of the mass without orchiectomy is the standard treatment for paratesticular masses, but more radical surgery is performed for the scrotal lesions, which cannot be separated from the testis. Following complete resection, tumour recurrence is rare in this location.¹¹ Conservative management with antibiotics, steroids and non-steroidal anti-inflammatory drugs, or observation with intense follow-up has also been described for unresectable masses on CT scan or laparoscopic examination.¹² Successful treatment of recurrent disease with various chemotherapeutic agents, including combination of vincristine, ifosfamide, doxorubicin and celecoxib, has been reported in some case reports.¹³ Impressive response to the ALK inhibitor crizotinib in a patient with ALK translocation has also been reported.¹⁴ It has been postulated that tumour locations have a prognostic influence on IMTs.^{15 16} The most favourable prognosis was noted for hepatic lesions, while abdominopelvic and retroperitoneal tumours show more aggressive behaviours with a poor prognosis.^{15 16}

Contributors SKS was responsible for manuscript writing, editing and research. SP was responsible for manuscript editing and research and patient follow-up.

Competing interests None declared.

Patient consent Consent obtained from guardian.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

Learning points

- ▶ Inflammatory myofibroblastic tumour (IMT) in the spermatic cord associated with undescended testes is rare.
- ▶ IMTs have clinical importance because they can be locally aggressive and can have metastases similar to a malignant lesion.
- ▶ Although genitourinary IMTs are rare in infants with undescended testes, they should be considered in differential diagnoses and strongly suspected on radiological imaging as they can be successfully treated by complete surgical resection or with conservative management.

REFERENCES

1. Umiker WO, Iverson L. Postinflammatory tumors of the lung; report of four cases simulating xanthoma, fibroma, or plasma cell tumor. *J Thorac Surg* 1954;28:55–63.
2. Cook JR, Dehner LP, Collins MH, et al. Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. *Am J Surg Pathol* 2001;25:1364–71.
3. Yee CH, To KF, Hou SM, et al. Inflammatory myofibroblastic tumor of spermatic cord in undescended testis. *Urology* 2009;73:1423.e9–1423.e12.
4. Swain RS, Tihan T, Horvai AE, et al. Inflammatory myofibroblastic tumor of the central nervous system and its relationship to inflammatory pseudotumor. *Hum Pathol* 2008;39:410–9.
5. Coffin CM, Watterson J, Priest JR, et al. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol* 1995;19:859–72.
6. Narla LD, Newman B, Spottswood SS, et al. Inflammatory pseudotumor. *Radiographics* 2003;23:719–29.
7. White JE, Chase CW, Kelley JE, et al. Inflammatory pseudotumor of the liver associated with extrahepatic infection. *South Med J* 1997;90:23–9.
8. Pungpapong S, Geiger XJ, Raimondo M. Inflammatory myofibroblastic tumor presenting as a pancreatic mass: a case report and review of the literature. *JOP* 2004;5:360–7.
9. Park SB, Cho KS, Kim JK, et al. Inflammatory pseudotumor (myoblastic tumor) of the genitourinary tract. *AJR Am J Roentgenol* 2008;191:1255–62.
10. Lim JH, Lee JH. Inflammatory pseudotumor of the liver. ultrasound and CT features. *Clin Imaging* 1995;19:43–6.
11. Patnana M, Sevrukov AB, Elsayes KM, et al. Inflammatory pseudotumor: the great mimicker. *AJR Am J Roentgenol* 2012;198:W217–W227.
12. Zhao JJ, Ling JQ, Fang Y, et al. Intra-abdominal inflammatory myofibroblastic tumor: spontaneous regression. *World J Gastroenterol* 2014;20:13625–31.
13. Johnson K, Notrica DM, Carpentieri D, et al. Successful treatment of recurrent pediatric inflammatory myofibroblastic tumor in a single patient with a novel chemotherapeutic regimen containing celecoxib. *J Pediatr Hematol Oncol* 2013;35:414–6.
14. Butrynski JE, D'Adamo DR, Hornick JL, et al. Crizotinib in ALK-rearranged inflammatory myofibroblastic tumor. *N Engl J Med* 2010;363:1727–33.
15. Lopez-Tomassetti Fernandez EM, Luis HD, Malagon AM, et al. Recurrence of inflammatory pseudotumor in the distal bile duct: lessons learned from a single case and reported cases. *World J Gastroenterol* 2006;12:3938–43.
16. Coffin CM, Hornick JL, 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.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow