Case report - Thoracic oncologic

Inflammatory myofibroblastic tumour at the pacemaker site

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Abstract

Inflammatory myofibroblastic tumour (IMT) or inflammatory pseudotumour is a histologically distinctive lesion occurring primarily in the viscera and soft tissue of children and young adults. We report an unusual case of IMT which had undergone malignant transformation in the chest wall at the pacemaker site. A 64-year-old male presented with a history of high fever, loss of appetite and weight loss of three months duration. He had a dual chamber pacemaker reinserted in the left infraclavicular region in the previous year. This was followed by a gradually enlarging hard swelling at the insertion site. The CT-scan showed a soft tissue mass encasing the pacing box, without intrathoracic extension. The trucut biopsy was suspicious of soft tissue sarcoma. A well encapsulated hard mass, with pacemaker embedded within it was resected en-bloc ensuring wide resection margins. Histology revealed fascicles of spindle cell proliferation with prominent inflammatory component, occasional spindle cells with prominent nucleoli and scattered atypical mitotic figures, with areas of focal necrosis. The lesional cells were negative for CD21, smooth muscle actin, ckit, cytokeratins and anaplastic lymphoma kinase 1. A diagnosis of IMT with malignant transformation i.e. inflammatory fibrosarcoma was made. He had adjuvant radiotherapy and uneventful recovery.

Keywords: Pacemaker; Inflammatory myofibroblastic tumour

1. Introduction

Inflammatory myofibroblastic tumour (IMT) or inflammatory pseudotumour is a distinct entity with characteristic clinical, pathological and molecular features. It has predilection for children and young adults and is associated with systemic symptoms in a minority of patients [1]. The most common anatomical locations are the abdominopelvic region, lung, and retroperitoneum, but virtually any site may be involved, including the somatic soft tissues, bone, larynx, uterus and central nervous system [2]. We report an unusual case of development of IMT at a permanent pacemaker insertion site.

2. Presentation

A 64-year-old male presented with a history of cough, high grade fever, night sweats, lethargy, loss of appetite and significant loss of weight of three months duration. He had a permanent pacemaker inserted for Stokes–Adams syndrome about ten years previously. A year before current presentation, this was replaced with a dual chamber pacemaker in the left infracavicular region. A small swelling had developed after the procedure at the pacemaker box site and was presumed to be an organising haematoma. However, the swelling gradually increased in size and attained the size of a cricket ball (Fig. 1a). On local examination it was round, smooth, non-tender and firm in consistency. It was fixed to the underlying muscle. The investigations revealed anaemia (Hb – 6.1 g/dl), increased inflammatory markers with ESR and CRP values at 35 mm/h and 155 mg/l, respectively. Repeated blood cultures did not grow any organisms. The transoesophageal echocardiogram did not show any evidence of endocarditis. The gastrooesophagoscop and colonoscopy were negative for any pathology in the gut to account for severe anaemia. He was reviewed by the haematologist and underwent a bone marrow biopsy which was normal. The CT-scan of thorax demonstrated 8–9 cm rounded mass in the left clavicular region superficial to the left pectoral muscle (Fig. 1b). It encased the pacemaker at its superior surface. The fine needle aspiration of the mass was suggestive of proliferative Fasciitis and raised a suspicion of sarcoma. In view of this, it was decided to excise the mass for diagnostic and therapeutic reasons. As a preparation for the procedure, the patient’s general condition was optimised and a new pacemaker was inserted in the opposite infracavicular region.

The surgical procedure was performed under general anaesthesia. The mass was well encapsulated with the pacemaker and its wires embedded within it. It was adherent to the pectoralis major muscle with no obvious infiltration into the chest wall. This mass with overlying skin and subcutaneous tissue, along with pacing box and pacing wire sheaths were removed en-bloc, ensuring wide resection margins (Fig. 2a).
The histological examination showed compact fascicles of spindle cell proliferation with prominent inflammatory component composed of lymphocytes, plasma cells and eosinophils (Fig. 2b). Numerous foci of necrosis were present within the specimen. The spindle cells reacted strongly and diffusely with a CD-34, and focally with EMA, but were negative with pancytokeratin, smooth actin, desmin, S100 protein, ckit, anaplastic lymphoma kinase (ALK) and bcl2. In view of the atypical mitotic figures and necrosis a histological diagnosis of malignant IMT was made.

The patient had uneventful postoperative recovery. His constitutional symptoms disappeared completely, anaemia resolved, the appetite returned, weight improved and the inflammatory markers returned to normal. The patient was discussed in local sarcomas related multidisciplinary meeting and a decision was made to offer him local adjuvant radiotherapy.

3. Comment

IMTs or inflammatory pseudotumours were first described in the lung where it was considered a reparative postinflammatory condition [3]. Similar lesions were later reported at extrapulmonary sites with predilection for children and young adults and associated with systemic symptoms in some patients [1]. Initially, these lesions were considered benign in nature, but in the 1990s Meis and Enzinger published a series of 38 cases of IMTs with significantly aggressive behaviour, including recurrences, metastasis and
death from the disease process [4]. The IMT or inflammatory pseudotumour is now considered a neoplasm with distinctive clinical, pathological and morphological features [5].

The patients generally present with a mass and symptoms pertaining to the location of disease, like cough, chest pain or rarely haemoptysis for pulmonary tumours and vague abdominal pain for intraabdominal lesions [5, 6]. IMTs may sometimes manifest as locally aggressive and destructive neoplasms. A constitutional syndrome consisting of fever, weight loss and malaise is seen in 15–30% of the patients [2]. The laboratory investigations may reveal microcytic anaemia, a raised ESR, thrombocytosis or polyclonal hyper-gammaglobulinemia. In some cases, the mass may only be detected after extensive work-up for these symptoms [5]. Preoperative diagnosis is rarely made in this condition as small biopsied specimens are insufficient for diagnosis because of the predominance of inflammatory cells.

The key histological feature is spindle cell proliferation in a myxoid to collagenous stroma with an inflammatory infiltrate composed primarily of plasma cells and lymphocytes. By ultrastructural analysis, IMTs are composed predominantly of myofibroblasts with a smaller fibroblastic component [7].

At molecular level rearrangements involving ALK locus on chromosome 2p23 has been noted in ILT giving further credence to neoplastic nature of these lesions. Over-expression of ALK protein can be detected by immunohistochemistry in half of IMTs, but is uncommon in older patients [7].

Surgical resection is necessary both to establish the diagnosis and to achieve surgical resection. Complete resection with clear resection margins leads to excellent outcome [8]. Incomplete resections have led to local recurrence [9], and hence combined modality therapy has been advocated for the management of these lesions [10]. The systemic manifestations and laboratory abnormalities are known to resolve after surgical resection, as was seen in the case described [2, 6].

Reviewing the English medical literature we have not come across any association of pacemaker and malignant proliferation at the site of insertion. As IMTs are thought to result from injuries traditionally, we can only presume that possibly an initial insult was the pacemaker placement and inflammatory response to the box which resulted in an IMT. To conclude, IMT is a distinctive neoplasm of intermediate malignant potential and is a diagnosis of exclusion in middle aged or older adults.

References


