Chest wall resection and reconstruction for Rosai–Dorfman disease masquerading as a chest wall sarcoma

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ABSTRACT

Rosai–Dorfman disease (RDD) is a rare benign histiocytic proliferative disease that can present as a pseudotumour of soft tissue. We describe the first case of RDD of the chest wall and mediastinum, outlining management by means of chest wall resection and reconstruction.

KEYWORDS

Chest wall – Mediastinal tumour – Sarcoma

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Rosai–Dorfman disease (RDD) is a rare benign histiocytic proliferative disease that can present as a pseudotumour of soft tissue. We describe the first chest wall resection and reconstruction for RDD of the chest wall and mediastinum.

Case history

A 62-year-old woman presented with an anterior chest wall mass that had been growing over a period of 10 months with no associated pain, dyspnoea, weight loss, night sweats or symptoms of mass effect. Computed tomography showed a 58mm x 48mm soft tissue mass eroding through the manubrium and the mediastinum (Fig 1), highly suspicious for sarcoma. Positron emission tomography showed metabolic activity with a maximum standardised uptake value of 11.2.

Blood test results and tumour markers were all within normal range. An anterior mediastinotomy and incisional biopsy were performed. Histology showed a nodular, rather well circumscribed and unencapsulated infiltrate involving the chest wall and sternal bone. The initial diagnosis was RDD with differential diagnoses of Langerhans cell histiocytosis (LCH) and Erdheim–Chester disease (ECD).

Chest wall resection and reconstruction was undertaken on one-lung ventilation in the supine position. An incision was made over the existing right anterior mediastinotomy scar with curvilinear extension towards the midline, where it joined a traditional median sternotomy incision. The right pectoralis major, sternum, second and third ribs, and right upper lobe was invaded by tumour. Right hemimanubriosternal excision was performed. The anterior aspects of the first, second, third and fourth ribs, the medial end of the right clavicle, the right sternoclavicular joint and the apex of

the lung were removed *en bloc* with the tumour. The tumour was dissected off the pericardium and great vessels through a thin tissue plane.

An intercostal drain was placed apically, and the chest wall was reconstructed with a composite methyl methacrylate and Prolene® (Ethicon, Somerville, NJ, US) mesh (Fig 2). The mesh was anchored to the surrounding skeletal edge. The lateral half of the right clavicle was anchored to the sternum with a metal plate (Synthes, Solothurn, Switzerland) including locking screws. Our plastic surgery team mobilised a left pectoralis major rotation flap for soft tissue coverage. The patient's postoperative period was unremarkable and she was discharged on day 11.

Figure 1 Cross-sectional computed tomography and coronal three-dimensional reconstruction

Figure 2 Intraoperative photographs: before resection (A), after resection (B), after chest wall reconstruction (C) and resected specimen (D)

Postoperative histology demonstrated an infiltrative lesion with foamy histiocytes, lipid laden macrophages and emperipolesis. Together with positive immunohistochemical staining for CD68 and S100 in histiocyte-like cells, this supported our diagnosis of RDD. LCH and ECD were excluded by negative CD1a/CD207 staining and the absence of a *BRAF* V600E mutation.

Discussion

RDD is a rare monocyte/macrophage proliferative disorder of varied biological behaviour.¹ It presents with lymphadenopathy, weight loss, malaise and night sweats. Extranodal disease tends to present with symptoms secondary to mass enlargement. Only about 3% of the largest series of reported RDD cases presented as extranodal soft tissue masses and only 36 cases of extranodal soft tissue RDD have been documented since 1969. RDD of the chest or mediastinum has typically been reported in middle-aged patients.²

RDD occurs primarily in soft tissue. However, a small number of cases with bone involvement have been reported.³ ECD and LHC were the primary differential diagnoses in this case. Histological examination in both diseases demonstrates foamy histiocytes, lipid laden macrophages and Touton type giant cells, all of which can be seen in all three disease processes. Positivity for S100 favours a diagnosis of RDD as staining in ECD is usually weak. Emperipolesis is also typical for RDD.⁴

We proceeded with surgery owing to localised symptoms and mass enlargement with invasion towards the great vessels. A report of RRD involving the heart has been described. Major cardiovascular reconstruction was needed but no chest wall intervention was required.⁵ To our knowledge, this is the first surgical case for RDD of the chest wall.

Although RDD is classified histologically as a benign disease, its clinical course is more typical of a malignant process. As a result, surgical excision is typically offered to those patients amenable. The role of adjuvant therapy is not clearly defined and the long-term prognosis is still not known.¹

Conclusions

RDD is a rare condition with an unpredictable course. The natural history of the disease is still not fully understood. It is benign but also progressive and has the possibility to recur. There is risk to the patient's life either by direct tumour penetration into an organ or major blood vessel, or by the progression from extranodal disease to disseminated nodal involvement.

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