An unusual cause of lymphadenopathy in a child

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DESCRIPTION

A 2-year-old boy presented to the Paediatric out patient department (OPD) with complaints of bilateral multiple neck swellings since 1 year which were insidious in onset, painless and progressively increasing in size. There was no history of fever, weight loss, night sweats, cough or abdominal pain. No history of contact with tuberculosis was noted. On examination, his vitals were stable. Mild pallor was present. There were multiple submandibular, submental, posterior cervical and preauricular lymph nodes, soft, non-tender, non-matted, largest of which was right submandibular 2.5×2.5 cm. Right axillary lymph node was 1.5×1.5 cm. There was no icterus, cyanosis, clubbing or bleeding spots on skin. Rest of the systemic examination was unremarkable. Chest X-ray was suggestive of right hilar and right paratracheal lymphadenopathy (figure 1). CT of thorax was suggestive of multiple nodes in right paratracheal, bilateral hilar, prevascular space and subcarinal region. CT abdomen and pelvis was suggestive of multiple lobulated nodes in celiac axis, peri-pancreatic and peri-splenic region. Our provisional diagnosis was generalised lymphadenopathy to rule out lymphoma. Second possibility was disseminated tubercular lymphadenopathy which was less likely. All blood investigations were normal except for iron deficiency anaemia and Mantoux test was non-reactive. Direct fine needle aspiration done from left cervical lymph node showed features of sinus histiocytosis with massive lymphadenopathy suggestive of Rosai Dorfman Disease (RDD) (figure 2). Parents were explained regarding the

Figure 2 H&E stain image (10x), showing large histiocytes exhibiting emperipolesis.

benign and self-limiting nature of the disease. It was planned that if systemic symptoms occur or if sudden enlargement of nodes causing vital organ compression or dysfunction occur, steroids will need to be started. In the follow-up for last 2 years, lymph nodes of same size are persisting, child has no systemic symptoms and is gaining weight and height appropriately.

RDD is a disease of non-malignant histiocytes that infiltrate lymph nodes or extranodal tissues. The hallmark of the RDD cells is emperipolesis, in which phagocytosis of lymphocytes or erythrocytes occurs.^{1,2} RDD presents clinically as massive,

Figure 1 Frontal radiograph of chest showing right hilar and right paratracheal lymph nodes.

Learning points

- ▶ Rosai Dorfman disease (RDD) is also known as sinus histiocytosis with massive lymphadenopathy and it typically presents with fever, painless cervical lymphadenopathy and elevated inflammatory markers.
- ▶ The disease has a predilection for the lymph nodes in the head and neck, but it can also involve any extranodal site, commonly skin and soft tissue, central nervous system, and less commonly, the gastrointestinal tract.
- ▶ RDD can be differentiated from malignant disorders like lymphoma and Langerhans cell histiocytosis by histology and immunohistochemistry.
- ▶ RDD is usually self-limiting and resolves spontaneously, rarely requiring systemic therapy.

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painless cervical lymphadenopathy with fever associated with elevated inflammatory markers. The most common extranodal sites involved are the skin and central nervous system and rarely, the gastrointestinal tract. The mainstay of diagnosis is immunohistochemistry with cells showing immunopositivity for S100P and CD68 while CD1a is negative. Many patients do not require treatment. However, for symptomatic localised disease, surgical resection remains the mainstay of treatment. A few patients may require immunomodulatory and antineoplastic agents.³

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