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Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): a rare case of subglottic narrowing

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Abstract
A case of subglottic narrowing caused by an extranodal deposit of sinus histiocytosis in an eight-year-old child with Rosai-Dorfman disease is presented. The airway obstruction was initially managed with a tracheostomy, and then on confirmation of the diagnosis by lymph node biopsy, by excision of the extranodal deposit with a CO_2 bronchoscopic laser.

Introduction

Sinus histiocytosis with massive lymphadenopathy is a very rare condition which typically presents in the first or second decade of life with massive bilateral cervical lymph node enlargement associated with fever, a raised ESR and polyclonal hypergammaglobulinaemia. Other lymph node groups may be involved with or without cervical adenopathy and in over 25 per cent of cases the disease involves extranodal sites. The condition is relatively unaffected by treatment, but often resolves spontaneously although it may run a protracted course over many years and deaths have been reported in cases where there has been infiltration of vital organs (Foucar *et al.*, 1988; Rosai, 1989). We present an unusual case of a very rare condition which presented with stridor caused by an extranodal deposit of sinus histiocytosis in the subglottis.

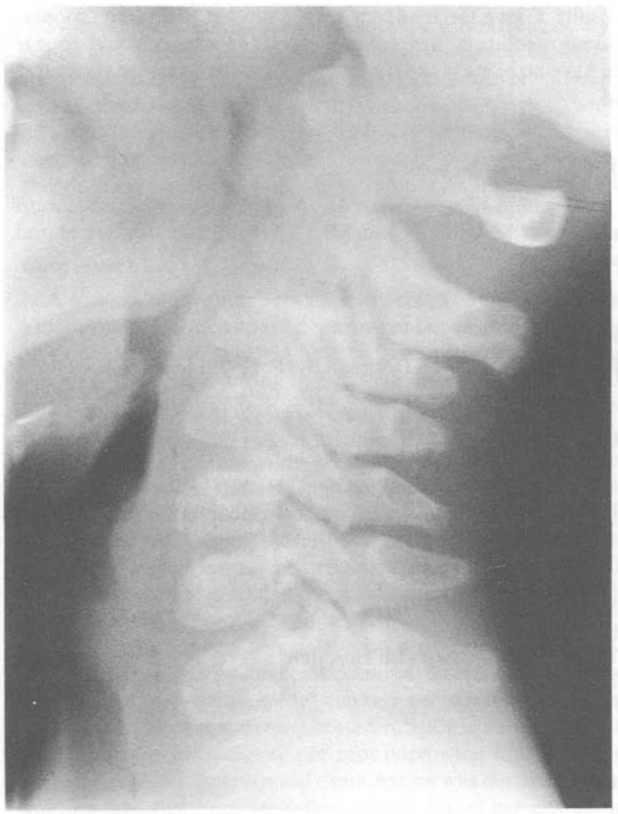

FIG. 1
Lateral X-ray of the neck showing a subglottic deposit of sinus histiocytosis.

Case report

An 8-year-old boy was referred to the Paediatric Department by his General Practitioner for investigation of cervical

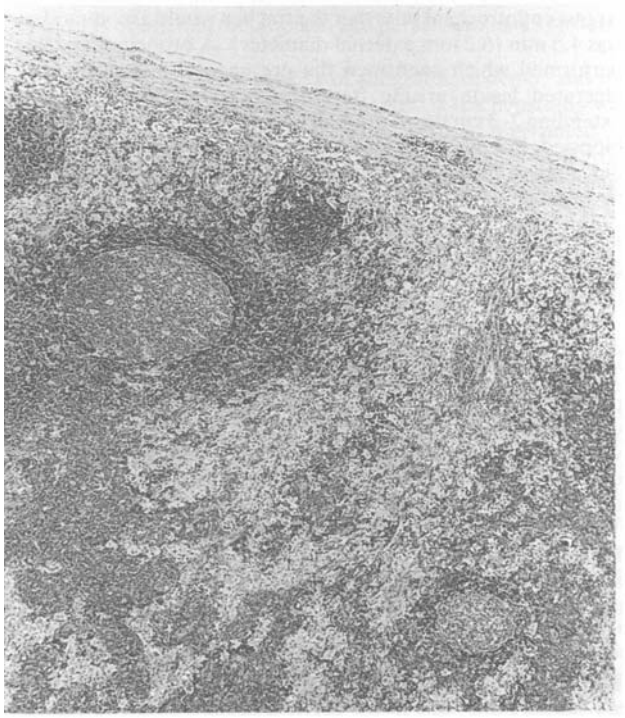

FIG.2
Section of the lymph node demonstrating normal architecture with hyperplastic follicles.

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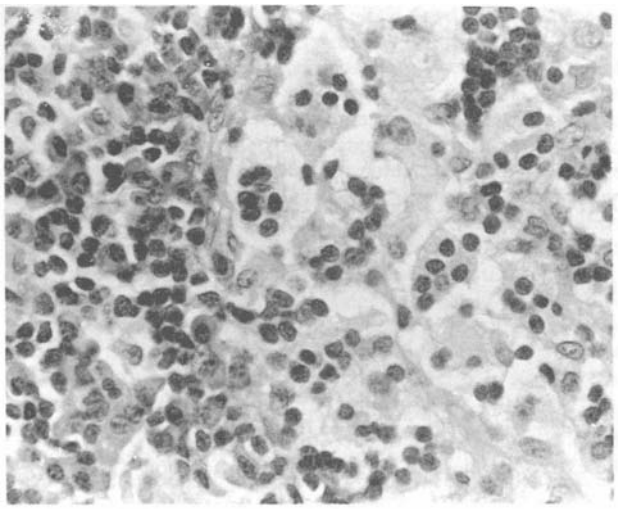

FIG. 3

Emperipolesis within histiocytes and numerous plasma cells in the paracortex.

lymphadenopathy which had been present for two months. He had bilateral cervical lymph nodes with no obvious focus of infection, but in the three weeks prior to being seen he had developed progressive dyspnoea. When he was seen in the Outpatient Department he had biphasic stridor and was admitted directly to the ward. Indirect laryngoscopy was performed which demonstrated a smooth subglottic swelling arising from the posterior wall of the trachea just below the level of the vocal cords. The lesion was also demonstrated on a lateral cervical neck X-ray (Fig. 1).

In view of his progressive airway obstruction, and the need to protect his airway during a diagnostic bronchoscopy, a tracheostomy was performed under general anaesthesia. The largest endotracheal tube that the trachea would accommodate was 4.5 mm (6.2 mm external diameter). A bronchoscopy was performed which confirmed the presence of a smooth, non-ulcerated lesion arising from the posterior subglottis and extending 2–3 cm down the wall of the trachea. The lesion was biopsied, but due to crush artefact this was not diagnostic, but did suggest that the lesion was inflammatory rather than neoplastic.

Ten days later a lymph node biopsy was undertaken and further biopsies taken from the subglottis. The cervical node was large, pale, soft and obviously adherent to other nodes by fibrous tissue.

Histology showed an enlarged lymph node measuring $4 \times 2.5 \times 2$ cm with a homogeneous pale brown, cut surface. The node showed mild capsular fibrosis. The architecture was preserved with hyperplastic follicles in the cortex (Fig. 2). Sinuses throughout the node were filled by large, pale histiocytes which showed emperipolesis and the adjacent paracortex and medulla contained numerous plasma cells (Fig. 3). The histiocytes showed strong cytoplasmic staining for S100 protein. The tracheal biopsy showed squamous metaplasia of the surface epithelium and in the underlying stroma, there was an aggregate of pale histiocytes, as seen in the node, which were surrounded by an infiltrate of lymphocytes and plasma cells. The features were those of Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy), with involvement of the tracheal mucosa.

Once the diagnosis was made it was decided that no specific systemic therapy was necessary; the patient was readmitted on two occasions and the remaining subglottic tissue excised using a CO_2 Laser. Three months after the initial presentation the patient was successfully decannulated and has remained well, though because of the nature of the disease long-term follow-up as an outpatient is planned.

Discussion

Rosai-Dorfman Disease is a rare condition. A case register has been established which, when last reviewed, contained 365 cases (Foucar *et al.*, 1988). Extra-nodal manifestations occur in approximately 30 per cent of cases. The majority affect the ear, nose and throat, predominantly the nasal cavities and salivary glands. Only one case of tracheal involvement has been reported (Foucar *et al.*, 1978). The case reported here demonstrated tracheal involvement severe enough to require a tracheostomy and was subsequently treated with local excision using a CO_2 laser to allow decannulation.

Diagnosis is made on the basis of histopathological appearances, and this case illustrates the diagnostic difficulties that may arise with biopsies of extra-nodal sites. The pathogenesis of the disease is not understood, and it is still not clear whether the characteristic histiocytic infiltrate represents the basic abnormality of represents a microscopically distinctive epiphenomenon (Foucar *et al.*, 1988). No consensus over management has been reached, but the use of antibiotics, radiotherapy and chemotherapy has produced at the best only transient results. Given the tendency for the disease to undergo spontaneous resolution, aggressive systemic therapies would appear to have no place in the treatment of localized disease (Foucar *et al.*, 1988).

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