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Rosai–Dorfman disease of the paranasal sinuses

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Abstract
A 57-year-old man presented with a history of nasal obstruction of five to six years duration. 'Nasal polyps' were removed on several occasions. He had previously had an episode of paraplegia which resolved after the removal of a spinal tumour. Histology from both sites was thought to represent a malignant fibrous histiocytoma. On presentation the patient had computed tomographical (CT) evidence of extensive ethmoidal disease, with threatened intracranial extension. He also had evidence of lung and retroperitoneal disease with pancreas and kidney involvement. The ethmoidal disease was considered potentially lethal and therefore a craniofacial resection was performed. Review of all the histology revealed that the diagnosis was extranodal Rosai–Dorfman disease (sinus histiocytosis). The patient's course is described, and the literature on this disease of unknown aetiology is reviewed.

Key words: Paranasal sinuses; Histiocytosis, sinus; Craniofacial resection

Introduction

Sinus histiocytosis is rare, and only one report of this disease affecting the paranasal sinuses was found (Wright and Richards, 1981), and one with respiratory tract involvement (Carpenter *et al.*, 1978). More recently, Tsang *et al.* (1992) have reported a case presenting as a persistent nodule in the ear.

The term 'sinus histiocytosis with massive lymphadenopathy' was introduced by Rosai and Dorfman (1969). This was a newly-recognized disease entity characterized by painless cervical lymphadenopathy, fever, leucocytosis, increased erythrocyte sedimentation rate and hypergammaglobulinaemia. A typical course is one of insidious onset, protracted duration of the active disease state and eventual spontaneous remission occasionally with subsequent recurrence. Lymph nodes other than those in the cervical area may be involved and extranodal involvement can occur. In our patient only extranodal involvement had occurred without any lymph node involvement.

Case report

A 57-year-old Caucasian male presented in February 1986 with a history of nasal obstruction of five to six years duration. During this period of time he had had a tumour removed from his right antral area on a number of occasions. The tumour was diagnosed as a malignant fibrous histiocytoma. Later on a lesion in his spine which caused temporary paraplegia was found to be of the same histopathology. On presentation the patient had tumour present in both ethmoids which had softened the cribriform plate and was extending down into the left antral area involving the sphenoid sinus and frontal sinuses (Figure 1 a–c). At the same time the patient was seen by a urologist who found that he had a fibrous mass lying in the left scrotal area and a plaque on the dorsum of the penis as well as an abnormally low kidney on the left side. Biopsy of the renal mass showed a similar histology. All of the histology from these areas and a review of the histology of the tumour previously removed in his antrum showed the same features. These were the features of extranodal sinus histiocytosis (Figure 2).

The patient was operated upon on the 2 April 1986. This comprized a craniofacial resection involving a left lateral rhinotomy, a right Lynch incision and a horizontal forehead incision. The craniofacial resection performed was carried out by the method described by Cheesman, Lund and Howard (1986) (see Figure 3). A 3 × 3 cm piece of bone was removed from the glabella area by means of an otological burr thus exposing the anterior cranial fossa and frontal sinuses. The posterior wall of the frontal sinus was removed exposing the anterior fossa dura. There was tumour to be seen in the floor of the frontal sinus which had quite extensively softened the posterior wall. The anterior fossa dura was stripped to the posterior edge of the jugum sphenoidale. Extensive tumour infiltration of the cribriform plate area and the floor of the anterior cranial fossa was already evident. Via the other two incisions, a left medial maxillectomy was performed with removal of all visible tumour in that area. Tumour was however attached to the orbital periosteum and to the dura. These structures were not removed but the tumour was stripped off as totally as possible. The anterior wall of the sphenoid sinus was opened, but there was no tumour in the sinus itself. The anterior fossa dura was reconstructed using fascia lata which was applied to it using Tisseel human fibrinogen glue (Immuno AG, Vienna). Skin from the left thigh area was also applied to the dura using the glue. The square of bone was replaced using interosseous wire.

The patient made an uneventful recovery and he is well without any local recurrence of tumour seven years after this procedure.

Discussion

Rosai and Dorfman (1969) first described sinus histiocytosis with massive lymphadenopathy. It is predominantly a disease of childhood and adolescence with 62 per cent of cases occurring in the first decade of life. Almost all patients present with substantial bilateral cervical lymphadenopathy. Other lymph node groups may also be involved in 78 per cent of cases. Additional symptoms include fever, weight loss, nasal discharge or obstruction and tonsillitis (Sanchez *et al.*, 1977). The disease almost invariably runs a benign course with gradual resolution of the lymphadenopathy, often over a period of many years. Involvement of extranodal tissues has been reported in 28 per

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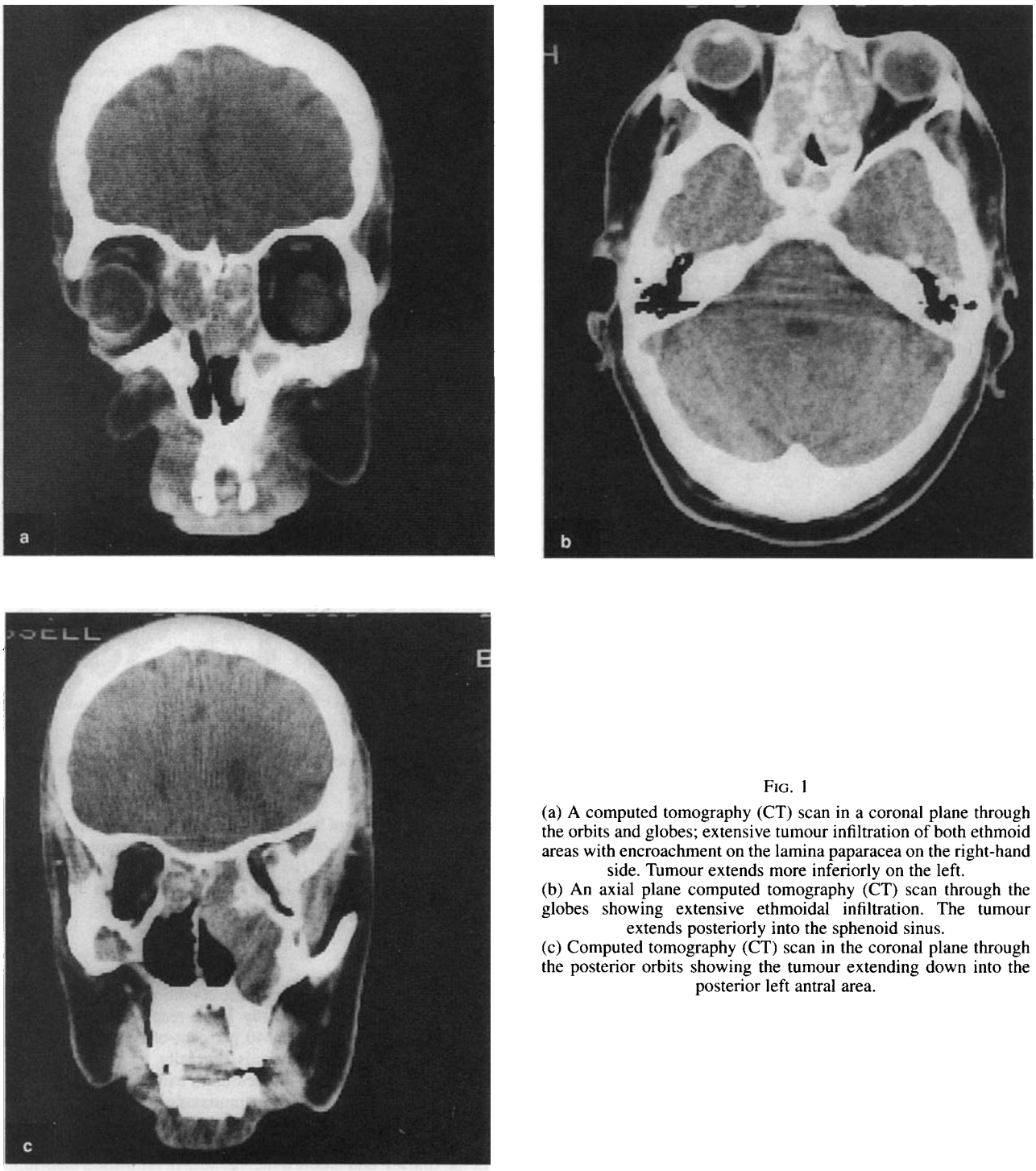

FIG. 1

(a) A computed tomography (CT) scan in a coronal plane through the orbits and globes; extensive tumour infiltration of both ethmoid areas with encroachment on the lamina paparacea on the right-hand side. Tumour extends more inferiorly on the left.
(b) An axial plane computed tomography (CT) scan through the globes showing extensive ethmoidal infiltration. The tumour extends posteriorly into the sphenoid sinus.
(c) Computed tomography (CT) scan in the coronal plane through the posterior orbits showing the tumour extending down into the posterior left antral area.

cent of cases with sinus histiocytocis and massive lymphadenopathy (SHML). These sites include orbit and eyelid, upper respiratory tract and salivary glands, skin and bone as well as testes. Sanchez *et al.* (1977) obtained follow-up information on 65 of the 117 cases. Five of these patients had died but only in one case was death directly attributable to SHML. Wright and Richards (1981) reported a patient with SHML which was initially involving the upper respiratory tract and which over a period of 17 years came to involve all lymph node groups with extension to lungs and kidneys ultimately resulting in uraemia and death.

This case is interesting in that it appears to be unique because it is the first in which extensive involvement of the lung and retroperitoneal tissues extending into pancreas and kidneys had been reported. Carpenter *et al.* (1978) reported one patient with respiratory tract involvement. Foucer *et al.* (1984) found 14 deaths in a register of 215 patients. Only in two cases was SHML infiltration clearly the cause of death.

The diagnosis of Rosai–Dorfman disease in a lymph node is usually straightforward, but in extranodal sites is often more difficult. The presence of dark-staining foci alternating with pale foci in areas is a characteristic low-magnification appearance found in most cases of SHML. A firm diagnosis can be made by identifying the distinctive histiocytes, which are typically immunoreactive for S-100 protein (Chan *et al.*, 1985; Eisen *et al.*, 1990). This immunostain also facilitates the search for lymphophagocytosis.

The patient in this report was interesting in that he had tumour extension into the retroperitoneal tissues, kidneys and penis as well as the paranasal sinuses, where recurrent tumour occurred after a number of removals. Softening of the cribriform plate had occurred and there was evident extension into the cranial cavity.

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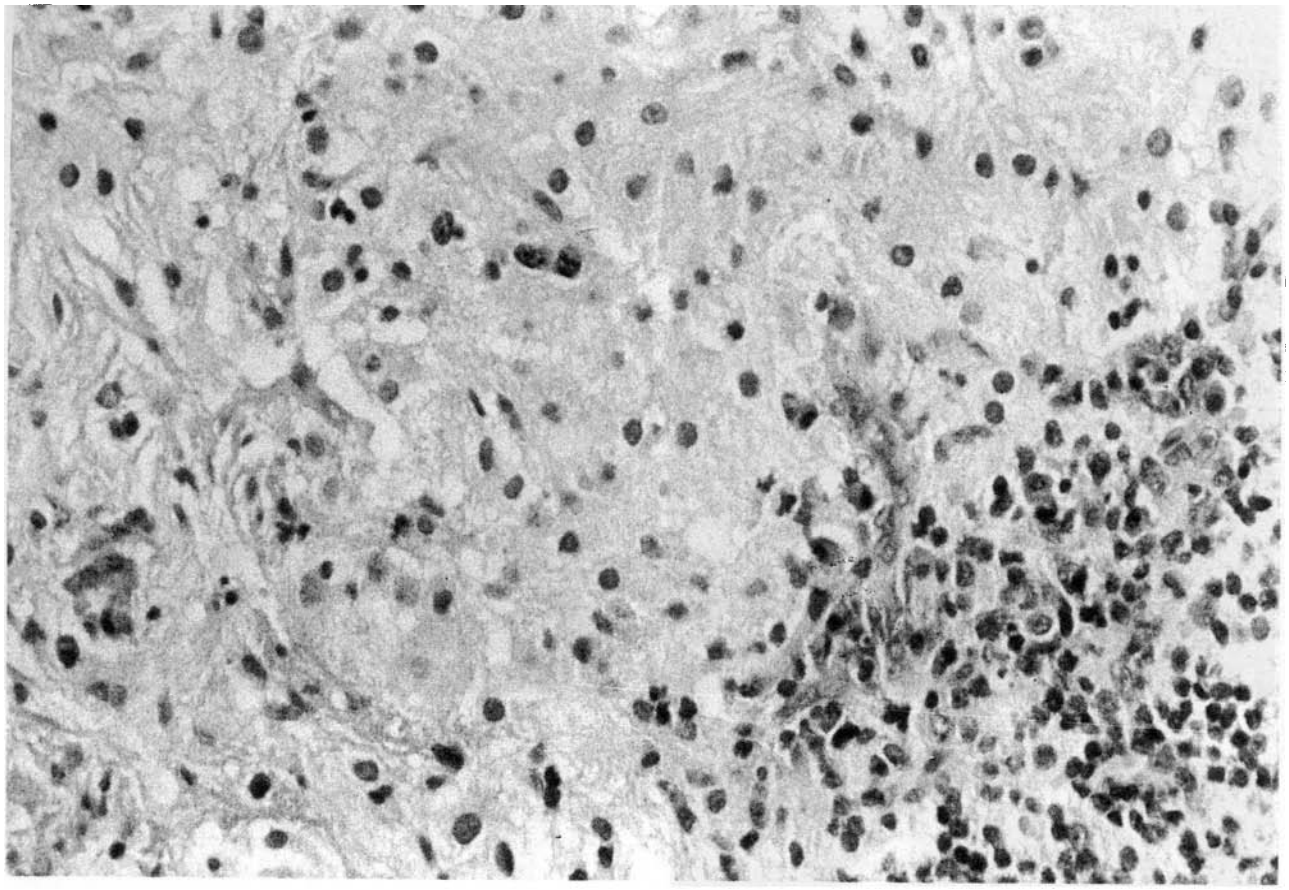

FIG. 2

Photomicrograph showing sheets of histiocytes and in one area a small aggregate of lymphocytes and plasma cells. The histiocytes exhibit minimal atypia of their nuclei (×400).

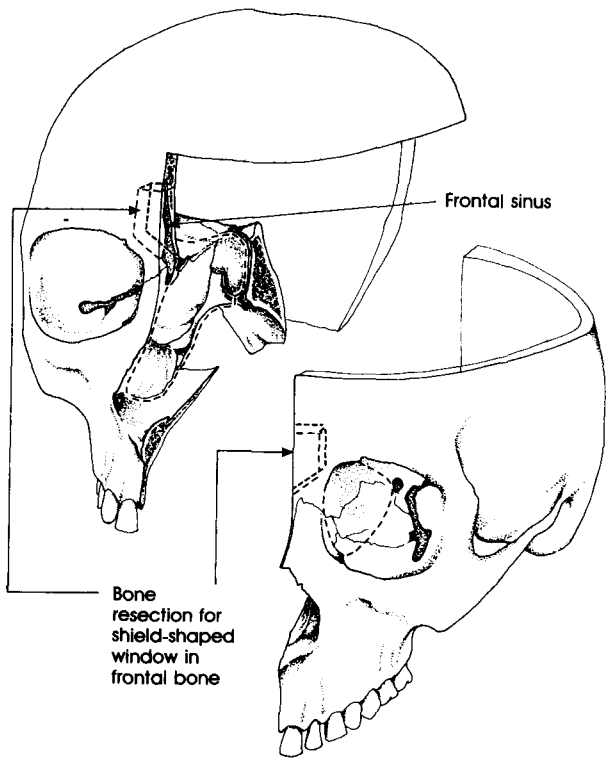

FIG. 3

An 'exploded' schematic representation of the cranio facial resection as performed in this patient. The dotted lines demonstrate the bony cuts and excisions made.

However this patient had no lymph node enlargement clinically. Spinal compression had also occurred, with paraplegia, which resolved after surgery. His disease was cleared by craniofacial resection, and there has been no local recurrence after seven years of follow-up.

Conclusion

Rosai–Dorfman disease is a rare condition usually associated with massive lymphadenopathy. It is a reactive histiocytic proliferative disorder which in 40 per cent of cases may involve extranodal sites. The course is normally insidious and self-limiting, but patients may succumb to extensive disease involving vital structures, associated immunological abnormalities or infection. We have reported our patient because of several unusual features; no lymph nodal involvement, rare involvement of the sinuses, potentially lethal because of cribriform plate infiltration, and hitherto unreported retroperitoneal disease with extension into the pancreas and kidneys. This patient demonstrates that in the case of vital structure extension, local removal can lead to resolution at these involved sites.

Acknowledgements

This patient was referred by Dr A. Hooper, of Durban, South Africa, and treated and operated upon at the Park Lane Clinic, Johannesburg, South Africa. Dr Ninnin works at the Lancet Laboratories, Jeppe str., Johannesburg, South Africa.

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