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Pathology in Focus

Coexistence of benign schwannoma and lymphoma in a nasal polyp

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Abstract
The existence of a combined benign schwannoma and lymphoma presenting as a nasal polyp has not been described in the English literature. We are reporting this rare combination in a 50-year-old male whose presenting symptoms were nasal obstruction, nasal deformity and headache. Examination of the left nasal cavity revealed a mass which was confined to the nose on computed tomography (CT) scan examination. Histopathology of the mass revealed a major component to be a benign schwannoma and a minor component a large B-cell lymphoma.

Key words: Nasal Polyps; Schwannoma; Lymphoma

Introduction

Benign schwannomas of the nose and paranasal sinuses are rare lesions presenting as only four per cent of head and neck tumours.[1] Nerve sheath tumours of the head and neck region mainly involve the VIIIth cranial nerve with only a small percentage occurring in the paranasal sinuses. The olfactory nerve or any of the smaller nerves arising from the nose and para-nasal sinuses may give rise to this tumour. Thirty-two benign schwannomas occurring within the paranasal sinuses were reported up to 1992.[2] Although these are benign, sometimes malignant degeneration and extra-cranial extension may complicate the course of a nasal schwannoma.[3]

Similarly the nasal region is an uncommon site for extra-nodal lymphomas composing about 0.5 per cent of all extra-nodal non-Hodgkin's lymphomas.[4] Their tendency to present with bulky disease, a high relapse rate and propensity to leptomeningeal spread distinctly differentiates them from other extra-nodal (Waldeyer's ring) and nodal non-Hodgkin's lymphomas in the head and neck region. Sino-nasal lymphomas are relatively rare in Western countries, in Asian populations they are the second most frequent group of extra-nodal lymphomas, after gastro-intestinal lymphomas.[5] The majority of these are of T-cell lineage.[6]

We describe in this report a very rare tumour in which both these lesions i.e. benign schwannoma and malignant lymphoma co-existed.

Case report

A 50-year-old labourer, a known drug addict, presented with a six-year history of left nasal obstruction, nasal deformity and headache. He further complained of facial swelling and rhinorrhoea. ENT examination showed mild nasal deformity with a grossly deviated nasal septum to the right side. The left nasal cavity was completely occupied by a fleshy, pale-coloured mass. The patient was evaluated by computerized tomographic (CT) scan of the head and neck which showed a 6×4 cm solid, homogenously enhancing mass in the left nasal cavity (Figure 1). There was bony

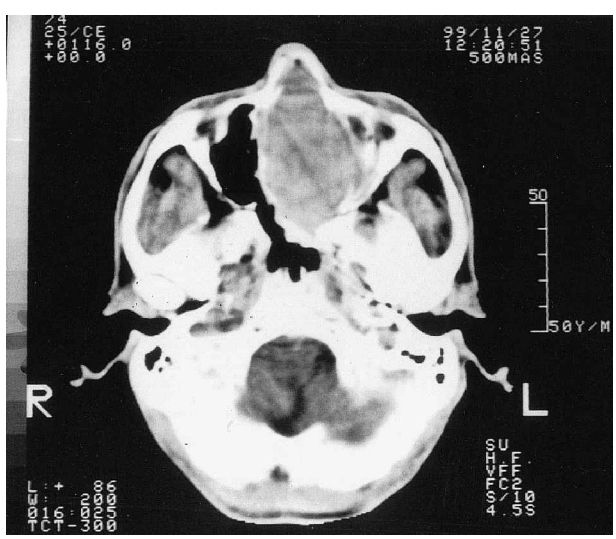

Fig. 1

CT scan shows left nasal cavity occupied by a homogenously enhancing mass. Impression was that of a benign lesion.

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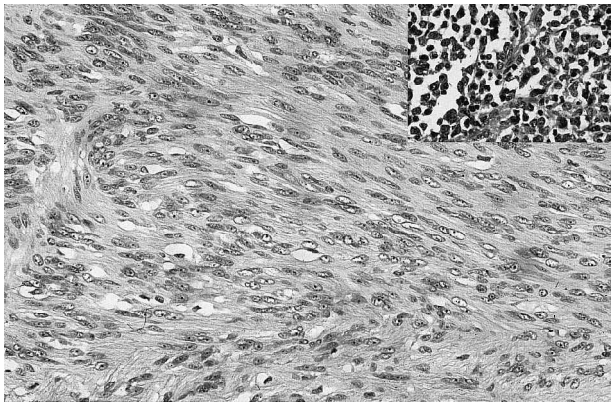

FIG. 2

A major component of benign schwannoma. Spindle cells were positive for S-100 protein (H & E; × 165). Inset shows a minor component of malignant lymphoma of B-cell lineage (H & E; × 200).

scalloping of the lateral and medial walls with no extension into the left maxillary sinus or orbit. Posteriorly, it was limited at the choanal opening. The tumour was completely removed by a lateral rhinotomy approach.

Gross features

Tissue was received, fixed in 10 per cent formalin which was then processed. It consisted of multiple yellowish brown fragments measuring 8.5 × 6 × 2.5 cm in maximum dimensions. 5 µm thin sections were cut. These were stained with haematoxylin and eosin. Sections were also stained with periodic acid Schiff for fungus.

Histology

Sections showed a lesion having two patterns of differentiation. There was a predominant component formed by cells with spindly nuclei having a variable degree of nuclear pleomorphism (Figure 2). Verocay bodies were also seen. However, no mitoses were seen in this component. A second focal component formed by a diffuse infiltrate of large lymphoid cells with atypical nuclei was also noted (Figure 2 inset). There were several mitotic figures. No lymphoid follicles or lympho-epithelial lesions were seen. Immunohistochemistry using the peroxidase-antiperoxidase technique showed spindly cells positive for S-100 protein antibody. Lymphoid cells were positive for the leukocyte common antigen (LCA) and Pan B marker (L26) but negative for Pan T marker (UCHL-1). According to the revised WHO Classification this was diagnosed as diffuse large B-cell non-Hodgkin's lymphoma (high grade). All antibodies were obtained from Dako Inco., Denmark. Parallel positive and negative controls were also run.

DNA extraction and EBV PCR analysis

DNA was extracted from a formalin-fixed paraffin biopsy from the area showing lymphoid infiltrate using a Nucleon genomic DNA extraction kit according to the manufacturer's instructions (Nucleon Bio-Sciences, Lanarkshire, UK). The purified DNA pellet was re-suspended in nuclease-free water and screened for EBV DNA using PCR primers for the gp220 region, as described by Feinmesser *et al.*[7] The amplification was performed on a Perkin Elmer thermal cycler model 9600 (Perkin Elmer, USA). A 10 µl aliquot of the amplified product was subjected to three per cent agarose gel electrophoresis. Specific products of 239 base pair size were detected with ethidium bromide staining and photographed.

Discussion

The differential diagnosis of nasal and para-nasal mass lesions include nasal polyps, mucocele, gliomas, papilloma, neuroblastoma, various sarcomas, carcinomas and lymphomas.[1]

A careful pre-operative examination of tumours is critical for identifying intra-cranial, intra-orbital or soft tissue involvement. Therefore, CT has become an important tool for the radiological evaluation of head and neck neoplasms. There was a clinical suspicion of a benign nasal polyp in our case. The CT examination revealed a solid homogenously enhancing mass in the left nasal cavity (Figure 1). The lesion was limited to the nasal cavity without extension into the left maxillary sinus or orbit. This further supported the clinical diagnosis of a benign lesion.

However, histological examination which was done on multiple blocks taken from the specimen revealed a predominantly spindle cell component without significantly enhanced mitotic activity. These areas were strongly labelled by antibody against S-100 protein. The unusual finding in one area was the presence of an infiltrate of atypical lymphoid cells, with large nuclei, exhibiting marked nuclear pleomorphism and atypia with prominent nucleoli. The lymphoid lineage was confirmed by immunolabelling with LCA and Pan B marker (L26). The specimen was further examined extensively revealing no further evidence of lymphoma.

In summary, we describe possibly for the first time in English literature a case of combined tumour showing a benign nerve sheath component along with a malignant component of a large cell lymphoma. The clinical and radiological examination was suggestive of a benign lesion but histological assessment revealed a focus of malignancy. This was seen in only one of the several blocks examined. It is prudent to extensively sample biopsy material submitted for histology. It also brings into question the reliability of diagnosis on a very small biopsy sample. This case had management and prognostic implications as the patient required combination chemo-radiotherapy for a large cell lymphoma that is likely to pursue a more aggressive course.

Epstein-Barr virus appears to be the stimulus for the development of non-Hodgkin's lymphoma.[8] The same cannot be said about the benign nerve sheath tumour as this association is not known to occur. A hypothetical sequence of events appears to be first appearances of schwannoma followed by EBV infection triggering a lympho-proliferative response. Most of the lymphomas of the paranasal sinuses are of T-cell origin, however our case showed a B-cell lineage. EBV-related lymphomas are commonly of T-cell phenotype. However, as in our case, a limited number of EBV-positive B-cell lymphomas have been described in the USA.[9]

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Dr I. N. Soomro takes responsibility for the integrity of the content of the paper.
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