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Case report

Infiltrative mass of the skull base and nasopharynx: A diagnostic conundrum

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HIGHLIGHTS

- Timely thorough investigation is critical to reduce the risk of irreversible damage.
- Prompt biopsy is essential to exclude both neoplasia and inflammatory conditions.
- Early corticosteroid administration is necessary to limit local infiltration.

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ABSTRACT

Inflammatory skull base masses are enigmatic and often behaviourally unpredictable. We present a case of idiopathic hypertrophic pachymeningitis (IHP) forming a central skull base mass to illustrate the process required when one investigates such skull base lesions. This is the first description of mass forming or tumefactive IHP extending into the nasopharynx. A 32-year old woman presented with frontal headaches and nasal discharge. She then deteriorated and was admitted with worsening headaches, serosanguinous nasal discharge and bilateral ophthalmoplegia. Multimodality imaging confirmed a destructive central skull base soft tissue mass involving the posterior clivus, floor of sphenoid sinus, nasopharynx and extending into both cavernous sinuses. Unfortunately, the patient continued to deteriorate despite treatment with broad-spectrum antibiotics. Cerebrospinal fluid, blood tests and transnasal biopsies for histology and microbiology did not reveal a diagnosis. Further neuroimaging revealed extension of the mass. Early corticosteroid treatment demonstrated radical improvement although an initial reducing regime resulted in significant rebound deterioration. She was stable on discharge with slowly reducing low dose oral prednisolone and azathioprine. We discuss the complexity of this case paying special attention to the process followed in order to arrive at a diagnosis of idiopathic hypertrophic pachymeningitis based on both the clinical progression and the detailed analysis of serial skull base imaging. Knowledge of the potential underlying aetiologies, characteristic radiological features, common pathogens and the impact on blood serology can narrow the potential differentials and may avoid the morbidity associated with extensive resective procedures.

1. Introduction

The investigation and management of skull base lesions often presents a challenge to the clinician. Inflammatory skull base masses are rare and unpredictable but can be associated with significant morbidity [1]. The differential diagnosis is extensive. Performing a diagnostic biopsy is often associated with risk of significant morbidity and hence the decision to opt for definitive or adjuvant surgical management further amplifies these risks. Knowledge of the potential underlying aetiologies, characteristic radiological features, common pathogens and the impact on blood serology can narrow the potential differentials and may avoid the morbidity associated with extensive resective procedures.

We present a case of idiopathic hypertrophic pachymeningitis (IHP) forming a central skull base mass to illustrate the process required when one investigates such lesions. This is the first description of mass forming IHP extending into the nasopharynx.

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2. Case presentation

A 32-year-old Nigerian woman presented to her primary care physician with a 1-month history of frontal headaches and serous nasal discharge. She was commenced on enteral antibiotics for presumed bacterial sinusitis. However, her symptoms progressed over the following 2 months. She was admitted to a tertiary care centre with worsening frontal headaches, ophthalmoplegia and serosanguinous nasal discharge. She had no other relevant past medical history. Examination demonstrated a right abducens palsy with associated mild right proptosis and ptosis.

Preliminary blood tests revealed a raised C-reactive protein (CRP) of 88 mg/L, erythrocyte sedimentation rate (ESR) of 128 mm/h, white cell count of 14.9×10^9/l, and neutrophils 12.8×10^9/l. Magnetic resonance imaging (MRI) confirmed an extensive destructive central skull base soft tissue mass involving the posterior clivus, floor of sphenoid sinus and nasopharynx. The mass extended into both cavernous sinuses, more marked on the right, with compression of both internal carotid arteries, for which she was started on prophylactic aspirin. The paranasal sinuses, orbits and brain parenchyma remained uninvolved. (See Fig. 1)

Causes of pachymeningitis were explored. Serum angiotensin converting enzyme (ACE), anti-nuclear antibodies (ANA), rheumatoid factor (RF) and anti-neutrophilic cytoplasmic antibodies (ANCA) levels were within normal limits and both her initial and repeat human immunodeficiency virus (HIV) test were negative. Cerebrospinal fluid (CSF) virology was negative, no organisms were grown on culture and protein was mildly raised at 0.48 mg/dL. She underwent two endoscopic transnasal biopsies of the lesion, both of which suggested reactive lymphoid hyperplasia alongside acute-on-chronic inflammation. This was thought to represent a possible acute response to infection. There were no features suggestive of lymphoma, granuloma or other malignancy. On advice from Infectious Disease a thorax/abdominal/pelvis CT was arranged, which did not reveal any other sites of inflammation.

She was commenced on intravenous meropenem 2 g three times daily, but continued to deteriorate with worsening right-sided ophthalmoplegia and ptosis. Microbiology and Neurology teams advised both anti-fungal and antituberculous treatment regimens along with prednisolone 50 mg once daily to minimise neurological morbidity. There was gradual improvement over the following two to three weeks with marked reduction in both ophthalmoplegia and ptosis. Her daily corticosteroids were slowly reduced, however this resulted in a dramatic deterioration in her visual acuity to perception of light in the left and finger counting in the right. Bilateral ophthalmoplegia recurred. Urgent MRI brain revealed progression of the mass to involve the pituitary fossa, further into both cavernous sinuses and increased compression of the internal carotid arteries. (See Fig. 2) She was commenced on high dose intravenous methylprednisolone. Within the first 24 h of therapy the patient noted a remarkable improvement in her visual acuity and her antibiotics were ceased.

An endoscopic transsphenoidal biopsy of sphenoid sinus mucosa and pituitary lesion was then undertaken. During the procedure thickened fibrotic dura was noted. Gram, Ziehl–Neelsen and periodic acid-Schiff staining were all negative. The histopathology excluded tuberculosis, sarcoidosis, granulomatosis with polyangitis and lymphoma. Immunoglobulin G4 (IgG4) serum levels were noted to be within normal limits excluding IgG4-related sclerosing disease. With these exhaustive investigations being negative, a diagnosis of idiopathic tumefactive hypertrophic pachymeningitis was proposed.

The patient continued to improve on the methylprednisolone and was converted to prednisolone 60 mg. Under supervision by the Neurology team she was commenced on azathioprine 100 mg, gradually increased to 200 mg once daily with a weaning course of corticosteroids.

At discharge she was stable on 200 mg azathioprine, 15 mg prednisolone and long-term low dose corticosteroids. The investigative MRI at this stage demonstrated significant reduction in uptake and size of the skull base lesion. Near vision had improved to N6 on the left and N8 on the right. (See Fig. 3) She is currently under close outpatient supervision and should she remain asymptomatic, a final MRI is planned for three years.

3. Discussion

Central skull base lesions extending into the nasopharynx are a rare occurrence. IHP is an uncommon condition characterised by diffuse or localised thickening of the dura mater (pachymeninges), causing progressive neurological deficits [2]. It is a diagnosis of exclusion. It is very uncommon for pachymeningeal hypertrophy to manifest as a large enhancing mass more accurately known as idiopathic tumefactive hypertrophic pachymeningitis (ITHP) [3]. We present an extremely rare case of ITHP extending into the nasopharynx, which has not previously been described in the literature. Our case highlights the importance of thorough diagnostic evaluation of any skull base or nasopharyngeal lesion to exclude infective, inflammatory, neoplastic and connective tissue aetiologies (see Table 1).

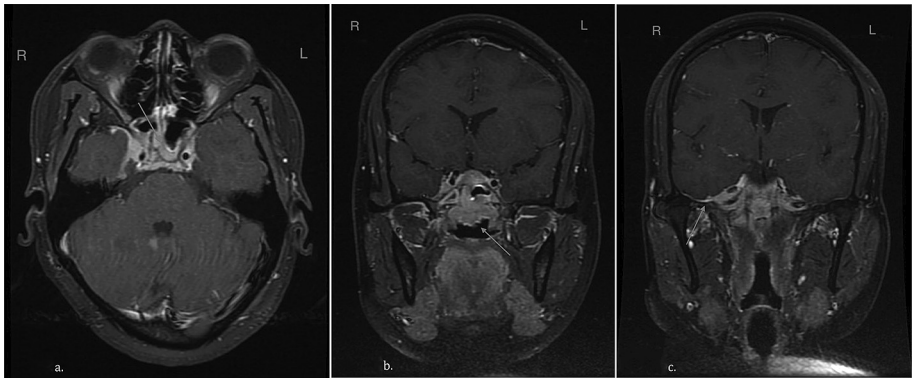

Fig. 1. a) MRI head T1 weighted post contrast axial slice. Demonstrating high uptake central skull base lesion at sphenoid sinus (arrow), cavernous sinus with compression of carotid arteries. b) MRI head T1 weighted post contrast coronal slice. A more anterior coronal slice demonstrating mass lesion at cavernous sinus, sphenoid sinus and extension into nasopharynx (arrow).c) MRI head T1 weighted post contrast coronal slice. A more posterior coronal slice demonstrating extension of mass into the right middle cranial fossa (arrow).