Extramedullary plasmacytoma of the frontal sinus secondary to multiple myeloma – a rare case of disease progression and relapse

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ABSTRACT

Background: Extramedullary plasmacytoma (EMP) is a rare disease involving the paranasal sinuses. It is an abnormal proliferation of monoclonal plasma cell in the extramedullary tissue and has tendency progressing into multiple myeloma. EMP accounts for 1% of head and neck malignancy and about 80% of cases with EMP occurs in the upper respiratory tract. Case Report: A 56-year-old lady, was diagnosed with multiple myeloma in which was treated with chemotherapy. Three-month post completion of chemotherapy, she developed rapid onset of painless left upper eyelid which was associated with diplopia and reduced vision. Clinical examination revealed a firm, non-tender, protruding mass on the left upper eyelid, extending from superior orbital rim towards lid margin. The vision of the left eye was to counting fingers. There were no associated nasal symptoms. Magnetic resonance imaging (MRI) of the brain and orbit reported a heterogeneous enhancing mass occupying the superior and lateral aspect of the extraconal region of the left orbit. There was evidence of cortical break at the inner wall of the left frontal sinus. The mass was extending into the left frontal sinus. Histopathological report of the tissue biopsy showed evidence of plasma cell myeloma with periorbital tissue involvement. Radiotherapy was commenced, and she responded well. Unfortunately, she developed relapse of her multiple myeloma and another relapse of EMP on her right forearm. For this, she has to undergo another cycle of chemio-radiotherapy. Conclusion: In view of its sensitivity to irradiation, EMP is best treated with radiotherapy. However, in the case presented in which there were disease progression and relapse, chemio-radiotherapy was instituted.

Spontaneous pseudomeningocele of sphenoid sinus or sphenoid mucocele? a diagnosis dilemma

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ABSTRACT

Introduction: Pseudomeningocele is an abnormal collection of cerebrospinal fluid (CSF) in the soft tissue that is not surrounded by arachnoid membranes. Any defects in the skull base can lead to spontaneous herniation of meningeal membranes and present as meningocele or meningoencephalocele. Bony defect may be small and clinically silent until a breach in the meninges appears, leading to a cerebrospinal fluid (CSF) leak. Typical presentations in anterior cranial fossa include CSF rhinorrhea and pulsatile or compressible nasal mass covered by attenuated meninges and mucosa. OBJECTIVE: To report a rare case of spontaneous pseudomeningocele of sphenoid sinus. Report: We describe a case of pseudomeningocele of sphenoid sinus in a 28-year-old gentleman. He presented to neuroendocrine with delayed puberty with diminished facial, axillary and genitalia hair, associated with headache. Otherwise, no rhinorrhea, nasal congestion or eye symptom was reported. No history of head trauma or base of skull surgery. Blood investigations showed hyperprolactinemia and hypotestosteronemia. Computed tomography (CT) scan revealed an expansible mass over bilateral sphenoid sinuses with complete erosion of intersinus septum and mass effect on the sella turcica and pituitary gland. Magnetic resonance imaging (MRI) features supported the diagnosis of sphenoid mucocle. Transnasal endoscopic sphenoidotomoy was performed to drain the mucocle. Intraoperatively, despite an anatomical puncture through the sphenoid ostium, alarmingly, the opening leaked out CSF. A dehiscent over left posterior wall was identified with a dural opening communicating with the left sphenoid sinus. The optic nerve and internal carotid artery were exposed. This was repaired with multilayer technique using fat, fascia lata graft, and nasal septal mucosal flap. Clinical improvement was observed post-operatively with no evidence of CSF leak and hypopituitarism. Conclusion: Pseudomeningoceles of base of skull are rare in the absence of trauma or iatrogenic injury. Surgeons should be alert to their presence as they can mimic a mucocle or nasal polyp.