

RAPID GROWING ESTHESIONEUROBLASTOMA (Case Report)

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SUMMARY:

Reported are the CT, clinical and histological features of a case of rapid growing esthesioneuroblastoma with intracranial extension. This tumour is most frequently reported to grow slowly inside the nasal cavities and can extend to the paranasal sinuses. The case investigated here occurred initially as an intracranial, retroorbital and intrapalpebral tumour which appeared suddenly one day and reached a significant size in ten days. It was diagnosed by computed tomography. Total resection combined with radiation therapy remains the most satisfactory treatment.

KEY WORDS:

Esthesioneuroblastoma, Nasal cavity neoplasms, Olfactory neuroblastoma.

INTRODUCTION:

Esthesioneuroblastoma, also called olfactory neuroblastoma, is a rare malignant tumour of the nasal cavities arising from the olfactory mucous membrane. Since the first report of this tumour in 1924 many cases of esthesioneurocytoma have been reported. The clinical symptoms are not specific, the most common presenting features are epistaxis, nasal obstruction and local pain (2). This tumour which can sometimes present with intracranial symptoms which is relatively slow growing and considered to be radiosensitive. Surgery followed by radiation therapy is the preferred treatment (7).

CASE REPORT:

A 67-year-old woman was admitted to hospital with an orbital mass. Her right eye was completely closed and exophthalmic. The orbital mass had taken 2 months to grow accompanied by a slight right-sided headache. She reported epistaxis starting 10 years ago and vanishing in the recent years. No symptoms associated with the nasal obstruction were

found. Neurological examination revealed complete anosmia. Computed tomography revealed a large soft tissue mass in front of the globe compressing it and invading the frontal and ethmoidal sinuses the destruction of the orbital roof and intracranial extension (Figure 1). CT obtained 2 months previously showed no sign of tumoral lesion (Figure 2). On operation an



Fig 1 : Preoperative Computerized Tomography scan.

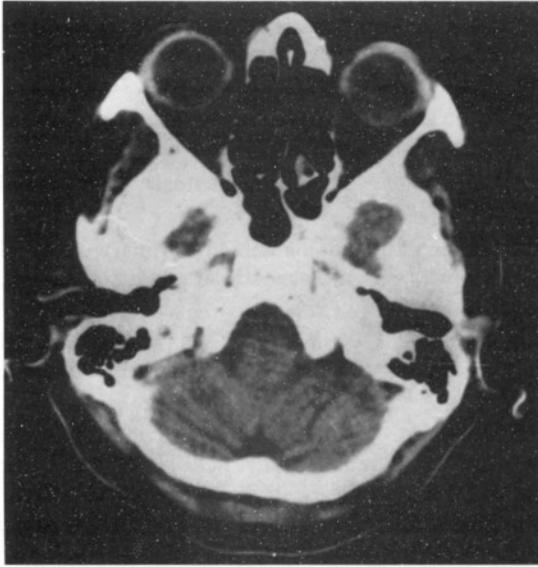


Fig 2 : Initial Computerized Tomography scan.

extradural approach was used and the tumour invading the frontal and ethmoid sinuses was resected. It was seen that the dura was infiltrated by tumoral lesion. Without a resection, the tumour was dissected from the dura and the dura was coagulated. The postoperative course was uneventful and surgery was followed by radiation therapy and chemotherapy. A follow-up CT scan obtained seven months later showed no tumoral lesion (Figure 3).



Fig 3 : Postoperative Computerized Tomography scan after radiation therapy.

DISCUSSION:

Olfactory neuroblastoma is a rare nasal cavity tumour (4) and was first reported by Berger and Luc in 1924 (10). This tumour has received notice principally in the journals of otolaryngology, radiology and pathology. The increasing recognition of olfactory neuroblastomas and their intracranial complications has become a matter of considerable interest among neurosurgeons. For the histogenesis of this tumour authors suggested olfactory mucosa, olfactory placode, sphenopalatine ganglion, the organ of Jacobson and vestigial paraganglia. Olfactory mucosal origin was widely accepted since both the olfactory mucosa and the tumour have nervous and epithelial components (7). The Hyams grading system for this tumour, from grade 1 to 4 based on the lobularity, mitotic activity, nuclear pleomorphism, fibrillary matrix, rosettes and necrosis, is becoming widely accepted (3,9). Our tumour grade was 2 according to this classification. Histopathological diagnosis of olfactory neuroblastoma is confirmed when the following elements are observed together: cords formed by small round cells, fibrillar intercellular matrix and rosettes (7).

CT and MRI scans show the tumour and its intracranial, intraorbital and sinus extension (5,10). Angiography is necessary to show vascularity and the potential for embolisation (7).

Intracranial extension usually occurs during the late phase of the illness and when occurs significantly worsens the prognosis. Esthesioneuroblastoma has been misdiagnosed clinically as encephalocele, meningioma, schwannoma, carcinoma and malignant lymphoma (10).

Extracranial metastases of esthesioneuroblastomas are relatively uncommon and the 5-year survival rate of patients without metastases or intracranial extension was reported as 50 % (7). There were no extracranial metastases in our patient.

Olfactory neuroblastoma is seen throughout life with the peak incidence between 10 and 39 years (8). As our patient was 67 years old, this was unusual.

Complete anosmia is suggestive of invasion of the cribriform plate. Severe frontal headache may be due to infection of the paranasal sinuses and does not necessarily signify neoplastic invasion of the sinuses or the intracranial cavity (8). Our patient had complete anosmia and headache when she was first examined. Although the CT scans obtained at that time showed no solid tumour but destruction of the lateral wall of ethmoid sinus and orbital wall.

The choice of treatment is surgery followed by radiation therapy (2,6), as this tumor is rather radiosensitive. During the course of craniotomy, it is wise to preserve the dura overlying the cribriform plate as a precaution against infection or rhinorrhea. Surgery must be performed in a multidisciplinary fashion and as a one step procedure.

Although olfactory neuroblastoma is known as a slow growing tumour (1,7) our case showed a rapid progression in two months and this pathology was confirmed with two follow-up CT scans.

CT scans obtained after radiation therapy showed no solid tumoral lesion.

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