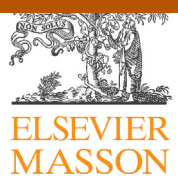
European Annals of Otorhinolaryngology, Head and Neck diseases (2014) 131, 49—51



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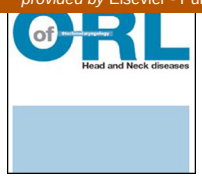

CASE REPORT

Leptomeningeal carcinomatosis from ethmoid sinus adenocarcinoma

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KEYWORDS
Adenocarcinoma;
Ethmoid sinus;
Paranasal sinus neoplasms;
Meningeal carcinomatosis

Summary
Introduction: Adenocarcinoma of the ethmoid is an aggressive tumor, with potential extension to surrounding structures. Leptomeningeal extension is a rarely reported entity.
Case report: A carpenter, aged 55, developed multifocal cranial nerve-related symptoms 1 week after resection of adenocarcinoma of the ethmoid, evolving towards deteriorated general health status and death 10 weeks later. Brain MRI showed diffuse contrast enhancement of the cranial nerves, and repeated cerebrospinal fluid (CSF) examination found increased protein concentration associated with decreased glucose concentration, without malignant cells. The diagnosis of carcinomatous meningitis was based on the association of clinical, CSF and brain MRI data.
Discussion/conclusion: Leptomeningeal dissemination of adenocarcinoma of the ethmoid is rare; diagnosis is guided by clinical signs. MRI reveals neurological spread, but the presence of malignant cells in the CSF is sufficient for diagnosis. Due to poor prognosis, the only currently available treatments are palliative.

Introduction

Primitive nasal adenocarcinoma is a rare tumor, usually located in the ethmoid [1,2]. Following various epidemiological studies implicating wood-dust exposure, nasal sinus adenocarcinoma was recognized as an occupational disease [3].

Ethmoid adenocarcinoma is locally aggressive, with risk of cerebral and meningeal extension. Leptomeningeal extension (also known as meningeal carcinomatosis or carcinomatous meningitis) consists in tumoral spread within the arachnoid, pia mater or cerebrospinal fluid (CSF) and is rare in ethmoid adenocarcinoma [4—10].

The present report concerns a patient presenting with meningeal dissemination following ethmoid adenocarcinoma surgery.

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doi:10.1016/j.anorl.2012.07.004

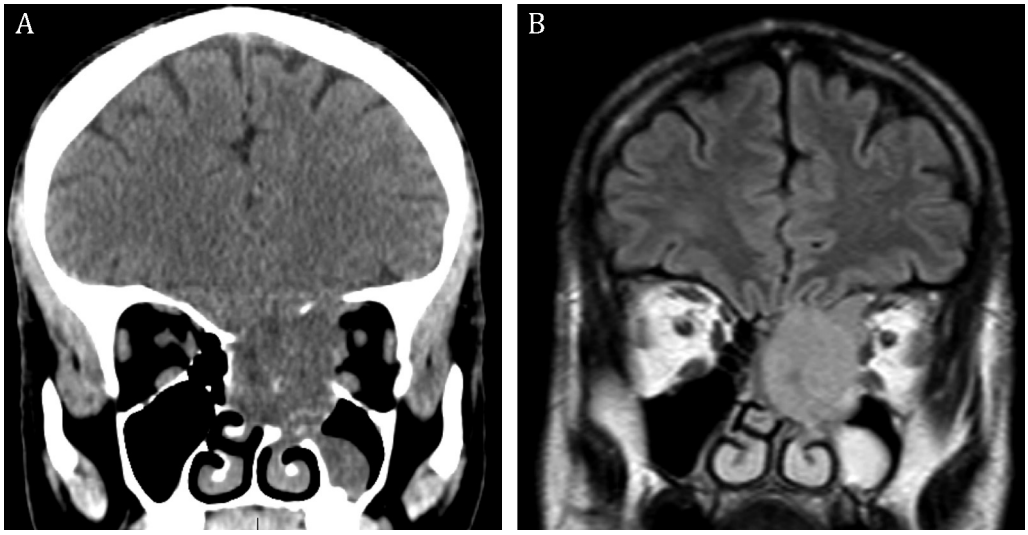

Figure 1 Facial imaging, coronal slices. A. Contrast-enhanced CT. Left ethmoid-orbital tumoral syndrome with destruction of the internal orbital wall and ethmoid roof. B. MRI FLAIR sequence. Left ethmoid-orbital tumoral syndrome with skull base invasion.

Case report

A 55-year-old man, who had worked as a carpenter for 37 years, with no particular medical history, was referred to our emergency unit for rapid loss of left-eye acuity. CT found a large tumor of ethmoid origin with left orbital extension compressing the optic nerve and destroying the left cribiform plate and ethmoid roof. Complementary MRI confirmed the tumoral aspect, with invasion of the base of the skull but no sign of intracerebral extension (Fig. 1). Anatomopathologic examination of a biopsy sample indicated mucinous intestinal-type adenocarcinoma with signet-ring cells. Resection was performed on a transfacial approach by a double neurosurgery-ENT team. There was macroscopic meningeal invasion with conserved cerebral parenchyma. Skull base reconstruction was performed with the technique developed in the Nantes Teaching Hospital since 1998, using the rectus abdominis muscular aponeurosis and abdominal fat held in place by a Silastic® sheet [11]. Histologic examination of the resection specimen found meningeal and focal cerebral parenchymatous infiltration.

Postoperative course featured sudden onset of right-ear hearing loss with secondary bilateralization, at 1 and 4 week respectively. Corticosteroids associated to vasodilators failed to provide improvement. A fall in right visual acuity occurred 18 days postoperatively; CT found no signs of optic nerve compression. Three lumbar punctures found increased protein concentration associated with decreased glucose concentration in the CSF, without malignant cells. Cerebral MRI found diffuse cranial nerve contrast uptake (Fig. 2).

The patient's general health status rapidly deteriorated, with onset of facial palsy, and impaired balance and swallowing, leading to death at 10 weeks postoperatively. Complementary postoperative radiation therapy was interrupted due to the general deterioration, and purely palliative treatment was pursued.

A diagnosis of carcinomatous meningitis was founded on the associated clinical, radiological and biological data.

Discussion

Overall survival in ethmoid adenocarcinoma (49% at 10 years) is significantly reduced in case of meningeal or

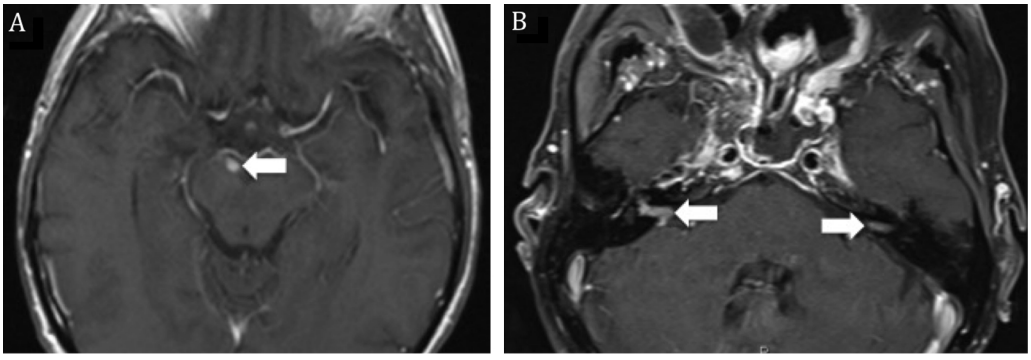

Figure 2 T-1 weighted gadolinium-enhanced cerebral MRI, axial slices. A. Right pre-mesencephalic uptake as of the 3rd cranial nerve (⇦). B. Bilateral uptake in acoustic-facial bundles (⇨⇦).

cerebral parenchymatous involvement [1]. Leptomeningeal spread from adenocarcinoma of the ethmoid has been rarely reported, although adenocarcinomas — basically mammary or pulmonary — are the solid tumors most likely to develop into carcinomatous meningitis [12]. Three of the eight cases reported in the literature revealed sinus tumor [4,6,8], despite five being secondary to tumor resection [5,7,9,10].

The dissemination pathways implicated in carcinomatous meningitis are 2-fold [12]: remote (via the hematogenic route or the peripheral nerves), or direct (direct contact with central nervous system, or leptomeningeal dissemination during surgical resection). In ethmoid adenocarcinoma with onset a few days after surgery, direct extension by dissemination during surgery is the more probable pathogenic hypothesis.

Diagnosis of meningeal carcinomatosis is founded on combined clinical, radiological and biological data.

Clinical signs concern tumor location, which is often multifocal, involving the cranial nerves (diplopia, impaired visual acuity, facial palsy, hypoacusis [13]) or spinal cord (equina cauda syndrome [4,5]).

Gadolinium-enhanced MRI has replaced CT, due to its greater sensitivity, in support of diagnosis. Images are non-specific, showing enhancement or presence of leptomeningeal, cranial nerve or spinal nodules, but may also be normal in 20% to 30% of cases [13].

Lumbar puncture provides positive diagnosis in case of tumor cells found in the CSF. Sensitivity is increased from 50% to 77% by iteration. Five percent of meningeal carcinomatoses show normal CSF [13]; associated non-specific abnormalities comprise high protein and low glucose levels in the CSF, with elevated CSF pressure in case of hydrocephalus.

Prognosis is poor. Median survival without and with treatment is respectively 4 to 6 weeks [12] and 8 to 16 weeks [13]. Treatment is palliative, associating intrathecal and systemic chemotherapy and radiation therapy. No decision tree has as yet been validated. As treatment is toxic and effective in only 20% of patients, selection factors for fairer prognosis have been investigated [13]. Among published cases, the two patients with lumbar location received external radiation therapy [4,5]; two received chemotherapy [5,7], resulting in clinical and radiological improvement in one case, nevertheless followed by disease progression and rapid death [7].

Factors of poor prognosis (meningeal and cerebral parenchymal infiltration [1], intestinal histologic type with signet-ring cells [3]) may partly explain the rapid clinical evolution and aggressiveness found in the present case, where surgery probably promoted meningeal dissemination of tumor cells.

Conclusion

Leptomeningeal dissemination of ethmoid adenocarcinoma is rare, but should be considered in case of suggestive semiology. MRI reveals the neurologic spread, but positive diagnosis is based on tumor cells found in the CSF. Whatever the treatment, prognosis is hopeless.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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