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Nasopharyngeal adenoid cystic carcinoma: a rare nasopharyngeal tumor

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Abstract. – Adenoid cystic carcinoma (ACC) is the most common histologic subtype of the salivary gland malignancies, but it is very rare in the nasopharynx with relatively few cases reported in the literature. We report a case of a 57 year-old woman with an ACC in the right Rosenmuller fossa of the nasopharynx, treated with combined radiotherapy and chemotherapy, without any recurrence or intracranial spread detected until the recent follow-up.

Key Words:

Adenoid cystic carcinoma, Nasopharynx, Radiotherapy, Chemotherapy.

Introduction

Previously termed "cylindroma" by Bilroth in 1856, the adenoid cystic carcinoma (ACC) of the nasopharynx is a rare disease with a slow evolution, but locally aggressive and thus prone to recurrence. Another important characteristic is its tendency to infiltrate neural structures and to spread perineurally[1-3].

Case Report

Ms. T.S.M., a 57 year-old Caucasian woman was admitted in our clinic for an undiagnosed 4-month initially lancinating pain of the right ear, which became continuous and more intense when lying down at the right side, and to which, a conductive hearing loss, tinnitus, ear fullness, paresthesias in the second division of the trigeminal nerve area and numbness of the right otic and parotid region were added within the next two months.

Previous consultations in other institutions did not reveal any pathology in the right ear and the pain was considered as a neuralgia of the second division of the trigeminal nerve, since the brain magnetic resonance imaging (MRI) performed showed no findings.

The analgesic therapy given had little effect on her pain. No remarkable personal history was found. The family history included her father died of multiple myeloma.

On physical examination a small mass in the right Rosenmuller fossa revealed from the endoscopy of the nasopharynx, an otitis media with effusion from the otomicroscopy and a decrease in sensitivity across the second division of the trigeminal nerve course from the neurologic exam. The remainder of the neurologic exam was normal. No cervical adenopathy was found.

Prior investigations had shown normal complete blood count, sedimentation rate and chest x-ray. The patient was submitted in a biopsy of the nasopharyngeal mass, under general anesthesia, and an MRI of the facial cranium, skull base and neck, which showed the extension and the limits of the lesion in the right lateral wall of the nasopharynx and the liquid content of the right mastoid cells (Figure 1). No cervical ganglions of important size and fortunately no intracranial spread of the tumor were found.

The histologic diagnosis revealed an ACC of the mainly cribriform model (grade II) (Figure 2). After discussing therapy modalities with the patient, she refused surgery and after the oncologic evaluation, the additional chest and abdomen MRI, and the bone scan, a combined radiotherapy and chemotherapy was proposed. Five days a week for 6 weeks and 2 days of the 7th week, the patient received a total dose of 6.5 Gy, combined with 68 mg cisplatin once a week for the same period. Therapy had minimum side effects.

After the end of the therapy the patient had again an imaging control showing no important

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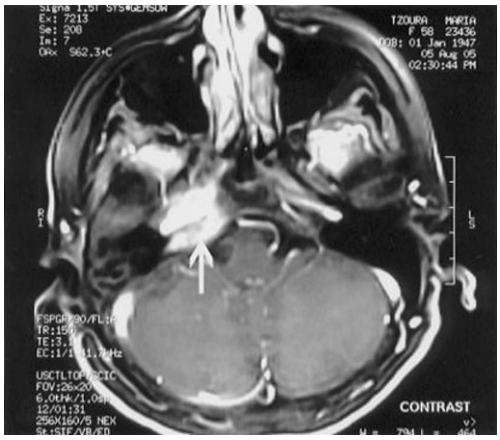

Figure 1. The MRI image showing the lesion and its limits. No intracranial spread was detected at the time of diagnosis.

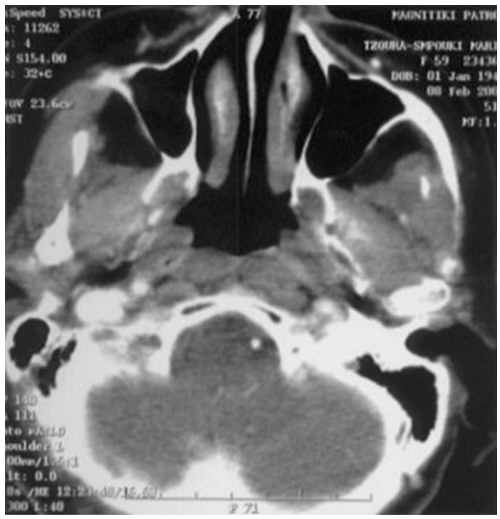

Figure 3. Post-therapeutic CT image: no lesion is shown on axial CT image.

findings in the nasopharyngeal region (Figure 3), and a physical exam revealing nothing but the post-radiotherapy mucositis.

Discussion

ACC is a malignant epithelial tumor of the salivary glands, occurring most frequently in the 5th decade of life, without sex predilection. It is the most common malignancy of the submandibular and minor salivary glands and it can be seen in other glandular tissues such as the lacrimal glands, ceruminous glands of the external auditory canal, esophagus, breast, prostate, uter cervix and Bartholin's glands. It accounts for less than 1% of all head and neck cancers, less than 10% of all salivary gland neoplasms but it is about 40% of all malignancies of the salivary glands[1,2]. The oral cavity is the most common site followed by the parotis, submandibular gland, paranasal sinuses, larynx, trachea and pharynx, in frequency order. In the nasopharynx, ACC can vary in presentation from an asymptomatic mass to severe ear and neurological symptoms, such as ear pain, fullness, tinnitus, glue ear, dizziness, hearing loss, facial pain, unilateral migraine-like headache, paresthesias, numbness, sensory loss across the trigeminal nerve course, abducens or oculomotor nerve deficits.

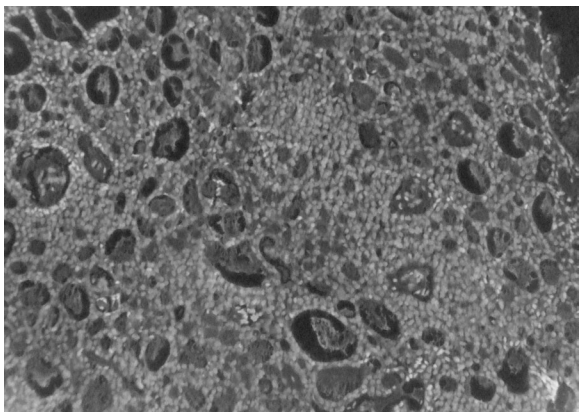

Figure 2. Histologic appearance of the tumor showing small round cells around the central cylinders, containing mucoid and hyaline material (stain: eosin-hematoxylin/magnification: 100 ×).

The histology of the tumor is interesting and characteristic. ACC appears as a "classic Swiss cheese", with uniform small round cells around the central cylinders and lamina, containing mucoid and hyaline material. The intercalated ducts of the salivary glands are thought to be their tissue of origin.

Three subtypes have been described by Perzin et al[4] in 1978, reflecting various degrees of progression of cellular differentiation, as well as aggressiveness of biologic behavior. The tubular subtype, which is the most differentiated form and the 2nd most frequent (30%), presents a recurrence rate more than 50% and an overall 9-year survival. The cribriform subtype, which is less differentiated than the tubular and the most common (50%) for most authors, presents a 90% recurrence and an 8-year overall survival. The

cribriform and tubular subtypes manifest a tendency for local infiltration. The less differentiated and thus the most malignant and aggressive is the solid subtype, accounting for 10% of the cases. It often gives distant metastases (70% overall) and mainly invades lung, brain and bones and has by far the poorest prognosis[5,6].

Lymph node metastases are rarer than distant (15% overall) and local recurrence rate is about 60% for the nasopharynx. An interesting point about the ACC is that in a specimen can be seen all 3 subtypes. On this basis, Szanto et al[7] in 1984 classified the ACC in 3 grades of malignancy: 1st grade represents the coexistence of tubular and cribriform model and the absence of the solid model, 2nd grade is mainly cribriform and less than 30% solid, and 3rd grade is mostly solid.

Another important issue, as mentioned above, is the tumor's tendency to invade the nerves and to propagate perineurally (80% of the specimens found invaded). Even if some studies show that there is not strong correlation between survival and perineural invasion alone, the majority believes that the invasion of a main nerve trunk or at least a large nerve by the tumor means a poor prognosis. Thus, its biologic behavior is rather deceptive, characterized by late and multiple reccurencies, but with a relative long survival with recurrent or distant disease[5,6].

The mainstay of therapy is surgical resection[8]. Today, skull base surgery facilitates total resection of advanced lesions. The main problem is the high local recurrence rate due to invasion of cranial nerves. The extend and the aggressiveness of the various surgical approaches which is needed in the nasopharyngeal ACC may cause hesitation to the patient, as in our case. Clearly, the best survival results are obtained through combined surgical and radiation therapy, as long as the surgical resection reaches at disease-free margins and preserves maximum physiologic function. Neutron radiotherapy[9] is applied in the nasopharyngeal ACC, because of its high local control rate for a long period of time (approximately 100% for a 5-year period in a 1993 study in the University of Washington). Chemotherapy has a limited role in the ACC of the nasopharynx and its use is still a matter of discussion. Cisplatin, 5-Fluorouracil, Doxorubicin and others are used in combination with radiotherapy, with reports of some success and remissions.

In conclusion, nasopharyngeal ACC is a rare tumor characterized by its tendency for local recurrence, perineural spread and distant metastases but with a relatively long survival in the presence of metastases. Its treatment is frustrating due to its deceptive biologic behavior. The combination of surgery and radiotherapy gives the best survival results with chemotherapy being still in controversy.

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