Cancer of the paranasal sinuses

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Cancers of the paranasal sinuses exhibit particular characteristics which distinguish them from other cancers of the upper aerodigestive tract. These characteristics include their great histological variability, epidemiology, the severity of a local disease, the rarity of metastases or lymphadenopathy and the infrequent presence of a second coexistent lesion.

Their development in natural anatomical cavities explains the often late diagnosis and fundamental role of the imaging techniques, either CT [6] or magnetic resonance imaging [5, 7, 10]. The severity of the disease is related to the local spread towards the skull base, brain, orbits and pterygomaxillary fossa.

Recent surgical advances, particularly techniques using a combined dual approach by the otolaryngologist and neurosurgeon, have provided access to extensive lesions and improved the terrible prognosis of intracranial spread. Bone involvement is unfortunately common and explains the low 5 year survival rate.

Clinical features

These are rare tumors representing only 0.2 to 0.8% of all cancers and 3 to 5% of cancers of the aerodigestive tract specifically.

80% of cancers occur in the ethmoidomaxillary sinuses, and pure ethmoidal and sphenoidal tumors are rare and lesions of the frontal sinuses exceptional.

The tumor predominantly affects men, rarely before the age of 40, the average age being between 40 and 70 years. Adenocarcinomas are common from the age 40, and carcinomas as well as adenoid cystic carcinomas are the rule in elderly patients between the ages of 50 and 70. Sarcomas are more often encountered in younger patients.

The etiologic role of infection or chronic inflammation is still controversial. There is certainly an associated chronic inflammation in the majority of epidermoid carcinomas, but its precise role, as for polyposis, is difficult to determine. Bucco-sinus fistulae of dental origin have also been implicated, but as yet there is no definite proof.

More certain are the malignant degeneration of papillomas and mostly the toxic effects of sawdust from hard or exotic woods which explains the increased incidence of ethmoidal adenocarcinomas in wood workers.

These cases are recognized as occupational diseases. Tannins in the sawdust are the responsible toxic agents. There is a long latent period of up to 30 years before the onset of these tumors. Other toxic industrial agents have been incriminated such as isopropanolol, asbestos, uranium and thorotrast. The histological appearances are diverse and are listed in order of frequency:

- epidermoid (60 to 65%), glandular or undifferentiated carcinomas (28%) [10],
- malignant lymphomas (10%),
- sarcomas of the soft tissues, bone or hemopoietic origin,
- melanomas,
- esthesio-neuroblastomas (neuroblastomas of the olfactory nerve) [20].
Carcinomas

These arise in the respiratory mucosa secondary to a metaplasia. Macroscopically the tumors are ulcerative, infiltrative, nodular or mixed and of a variable degree of differentiation. The undifferentiated form is the commonest and is associated with the highest incidence of lymph node involvement.

The adenocarcinomas are classified as well differentiated, papillary or trabecular, colloid or poorly differentiated adenocarcinomas.

The adenoid cystic carcinomas (cylindromas) are considered to be malignant tumors but are slow growing. They usually arise in the vault of the palate.

The sarcomas, hematosarcomas, fibro- or osteosarcomas, chondrosarcomas or angiosarcomas, depending on their cell of origin, have a very poor prognosis.

The malignant melanomas are rare and often achromatic, they frequently metastasize.

Esthesio-neuroblastomas are tumors which arise from the olfactory bulb. They are peculiar due to their localization on the cribriform plate and their early attachment to the anterior aspect of the skull base. They extend inferiorly towards the nasal fossae.

Other tumors are rare but gliomas, meningeomas, sympathomas, schwannomas, plasmacytomas and oncocyctic tumors may all be encountered [1]. The presenting signs are very variable, nasal symptoms predominate, and patients often present with recurrent minimal epistaxis, chronic mucopurulent rhinorrhoea and progressive nasal obstruction. Unilateral involvement is extremely suggestive.

Deformities occur later and indicate tumor extension to the bony walls of the sinus.

Neurological symptoms occur late and are due to invasion of the nerves with resultant anesthesia or paresthesia of the territory supplied by the nerve, particularly the maxillary nerves. Pain is often an indication of skull base involvement. The signs of orbital involvement are unilateral disease with diplopia, reduced visual acuity, lid edema and exophthalmos. These are only present in 50% of cases. The buccodental signs are not rare, with deformity or abnormal mobility of a tooth, poor healing following extraction or the formation of a fistula. Trismus is a later sign due to involvement of the pterygo-maxillary fossa and muscles of mastication. Lymphadenopathy is rarely the presenting feature.

Imaging techniques

The imaging is indicated only after a careful clinical examination, including a full neurological and general examination and endoscopy of the otorhinolaryngological system. Standard radiographs of the sinuses in the frontonasal, Blondeau (water), Hirtz and lateral projections may give clues to the diagnosis of fully developed tumors, but are totally insufficient for assessing tumor spread. Tomograms [35] have been