Cylindroma in the region of the Gasserian ganglion

Case report

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✓ A rare case of cylindroma of the region of the Gasserian ganglion is reported. Signs and symptoms simulated a trigeminal neurinoma. Total removal was attempted; however, even when it seems to have been feasible as in this case, it may not be so since what appears to be erosion of the base of the skull may in fact be infiltration.

Key Words • cylindroma • adenocarcinoma • brain tumor • Gasserian ganglion • skull tumor

Cylindromas, which were at first regarded as connective-tissue tumors, and later as epithelio-connective-tissue tumors, are now considered to be a peculiar variety of the so-called “mixed tumors of the seromucous glands” whose specific location is often in the salivary and lacrimal glands. On rare occasions they have been found in other sites, such as the breast, intestine, pancreas, vulva, the mucous glands of the bronchi, or the sweat glands of the skin. The tumor in the case we are reporting was in the region of the Gasserian ganglion.

Case Report

This 45-year-old woman was admitted to the hospital on August 31, 1969, with a 3-year history of nearly continuous pain in the right half of the face. Periodically the pain became severe, lasting a few minutes; it was refractory to all analgesic drugs. The attacks had become more frequent over the past 8 months. Occasionally she had dysesthesias (tingling, sensations of heat) in the same side of the face. Double vision had been present for 2 months.

Examination. General physical examination was negative. There was hypesthesia to touch, heat, and pain in the right trigeminal distribution, and a right abducens paresis. Chest x-ray films were normal. Skull films showed no clear outline of the tip of the right petrous bone; there was osteoporosis extending toward the base of the bone (Fig. 1) but no signs of intracranial hypertension. Right carotid and vertebral angiography by the right brachial route were normal.

Operation. Through a right frontal bone flap, the dura mater was opened and the temporal lobe raised, revealing that the dura of the base of the skull at the level of the Gasserian ganglion was infiltrated and elevated by a tumor that looked like an “en plaque” meningioma. The trigeminal nerve, which was firmly adherent to it, was cut and the mass removed apparently totally; most of the tumor, which was the size of a chestnut,
was outside the dura. The base of the skull, which was eroded, was coagulated at many points.

Postoperative Course. Recovery was uneventful. When discharged the patient had a right trigeminal anesthesia and right sixth nerve deficit.

Histologic Examination. The tissue of the tumor seemed to be composed of numerous clusters of oval epithelial cells of varying size, supported by abundant stroma (Fig. 2). The smaller clusters were compact; many of the larger ones had a central cavity filled with a mauve, amorphous striated substance. The epithelium of the larger cavities was made up of a single layer of cells, while that of the smaller cavities had two or more layers. The boundaries of the cytoplasm were often blurred. The nucleus was finely granular and stained well; there was little variation in the size or shape of the cells, and only a few mitoses. The diagnosis was cylindromatous epithelioma.

Discussion

The microscopic characteristics of a cylindroma are as follows. Submerged in a connective-tissue stroma lie roundish or elongated epithelial clusters of varying size, the majority having a cavity in the center filled with mucous-like colloid; these cavities, which give the tissue its typical sieve-like appearance recalling the architecture of the thyroid gland, are demarcated by elongated-oval epithelial cells of regular size, without nuclear anomalies, and with occasional karyokinetic figures. Although there is no clear histologic evidence for malignancy, the cylindroma is usually regarded as malignant, or potentially malignant,7,12 because of the very high local recurrence rate. Distant metastases are rare.
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Fig. 2. Upper Left: Photomicrograph showing typical pattern of a cylindroma: solid and cystic epithelial clusters supported by connective-tissue processes. H & E, ×100. Upper Right: Cystic formations filled with an eosinophilic mucous-like substance are seen bounded by epithelial cells with hyperchromatic nucleus. H & E, ×200. Lower Left: At some points the stromal component tends to prevail over actual tumor tissue. H & E, ×100. Lower Right: At high magnification the cells have a uniform appearance with a few mitoses (arrow). H & E, ×500.
Primary intracranial examples of this tumor have been reported. In 1964 Gonzalez and Zülich, who have devoted a great deal of attention to this tumor, reported nine cases and collected eight others from the literature. Some of the latter had been classified as sarcomas, endotheliomas, or neuromas, but Gonzalez and Zülich, going by the description and figures, considered "not always with great certainty" that they belonged to the "cylindromatous epithelioma" group.

The apparently primary intracranial cylindromas have been found in two regions of the base of the skull: the chiasmal region (four cases) and the Gasserian ganglion region (14 cases). Gonzalez and Zülich suggested that the tumors of the chiasmal region originate from the nasal fossae or paranasal sinuses and that they invade the skull via the cribiform plate, whereas those of the Gasserian ganglion region arise from the glandular epithelium of the eustachian tube which, like the mucosa of the nose and paranasal sinuses, derives from the epithelium of the primitive buccal cavity. In support of their hypothesis they point out that all tumors presenting regressive phenomena on the lines of those exhibited by cylindromatous tumors arise from bucconasal rests.

These cylindromas show similarities with the rare carcinomas (non-cylindromatous) of the nasopharynx or ear which penetrate the base of the skull, giving rise to a multiple cranial-nerve deficit pattern but to no other manifestations. In such cases the primary tumor may be so small as to escape notice throughout life and even at necropsy.

Cylindromas have developmental and clinical peculiarities that distinguish them from cylindromas of the juxtaocular regions (orbit, paranasal sinuses, parotid), in which the intracranial site is clearly secondary.

Our case does not differ from other cylindromas of the Gasserian ganglion region reported in the literature. Most of the previous cases of cylindroma were in the midline, at the level of the tip of the petrous bone, and usually in the middle cranial fossa, rarely toward the posterior fossa. The patients were mostly middle-aged women, as in our case. The clinical history lasted 6 years in one case, 3 years in two, 2 years in five, 1 year in three, and 8 mos in one. In our case the history was 3 years. Clinical trigeminal-nerve disturbances were present in all cases, with unilateral trigeminal pain in nine, and hypesthesia in three. No case presented trigeminal neuralgia and/or hypesthesia alone, and the same applied to our case, in which sixth cranial-nerve paresis was present. In all but three cases the trigeminal disturbances were accompanied by a homolateral deficit of the oculomotor nerves (paresis of the sixth nerve or ptosis or complete ophthalmoplegia) and in some cases deficits of the seventh, eighth, and twelfth nerves, choked disc or unilateral temporal pallor with papillary scotoma. In one of these cases the anterior and posterior growth of the tumor gave rise to a cerebellopontine syndrome associated with ipsilateral exophthalmos, choked disc, primary optic atrophy, and paresis of the contralateral sixth cranial nerve. In the three cases without oculomotor deficit the trigeminal pain was associated with a "marked decrease in visual acuity," or with hypesthesia and primary optic atrophy, or with ipsilateral facial paralysis. As in our case, skull films showed slight erosion of the tip of the petrous bone and of the sphenopetrous region.

In brief, cylindromas usually present signs typical of most well-known tumors of the region, such as those of neurinomas of the fifth cranial nerve or meningiomas of the trigeminal cavity. Ours and all the cases of Gonzalez and Zülich were operated on for trigeminal neurinoma. Cylindroma was not suspected at operation where the tumor seemed to have the macroscopical characters of a neurinoma, and alterations of the ganglion and roots seemed to continue into the tumor. Bone was consistently eroded and in some cases infiltrated but the dura mater was intact. In our case the tumor looked more like an "en plaque" meningioma than a neurinoma: the dura appeared to be infiltrated and the tumor had developed intradurally, though less than extradurally, with the trigeminal nerve firmly adhering to the tumor mass. The corresponding part of the base of the skull was clearly eroded but did not seem to be infiltrated, and apparently complete removal was effected with relative ease.

Chiasmal cylindromas have different symptoms including blindness, ptosis and exophthalmos, bitemporal hemianopsia, and temporal pallor of the optic disc. Skull films

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show erosion of the planum sphenoidale, sometimes with thinning and partial destruction of the sella.

Gonzalez and Zülch observed two local recurrences and one metastasis in the neighborhood of the tumor in the span of 1 to 6 years; they gave 2 to 6 years as the postoperative survival but admitted that this is of no statistical value because of the small number of known cases. It must be added also that in some intracranial cylindromas the changes of the base of the skull, judged to be infiltrative rather than erosive, had suggested that tumor removal had been incomplete. Eneroth, et al.,4 who studied numerous cases of cylindroma of the palate, have concluded that tumors presenting mainly as solid structures are less differentiated and biologically more malignant than those with mainly cystic structures; they also emphasized that cylindromas are highly radiosensitive.

Summary

We have reported the case of an apparently primary intracranial cylindroma of the Gasserian ganglion and have reviewed the related literature.

References


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