Intracranial Spread of Adamantinoma

Case Report

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Adamantinoma is a rare tumor of the jaw, occurring more commonly in the mandible than the maxillae. It is commonest in the third decade of life, is equally distributed between both sexes, and is said to be more common in the white races than in negroes. The tumor grows slowly over a period of many years, but is, however, locally malignant and invariably recurs unless adequate removal is undertaken. It is unusual for this neoplasm to spread to other regions. We are reporting a case of intracranial tumor, which proved to be an adamantinoma that had its origin in the mandible.

Case Report

The patient, a female Jamaican immigrant, had a painless swelling removed from the body of the left side of the mandible in Jamaica in 1948, when she was aged nineteen years. This swelling had then been present for at least 1 year. Histologically, it was reported to be an adamantinoma. The tumor recurred locally, and necessitated further surgery in 1949, 1954 and 1958, when it was found to have spread into the spheno-palatine fossa. The patient remained without symptoms until September, 1959, when she developed diplopia due to weakness of the left external rectus muscle. In June, 1960, she began to complain of frontal headaches which gradually became more severe, and which later were accompanied by nausea, vomiting, vertigo, and ataxia severe enough to prevent walking. Dysarthria and dysphagia quickly followed.

Examination in July, 1960, revealed a painless swelling, thought to be a recurrent tumor, on the mucous surface of the left cheek. There was paralysis of the left 5th, 6th, and 12th cranial nerves, with gross nystagmus, cerebellar ataxia and a right-sided spastic hemiparesis. There was no papilledema or other sign of raised intracranial pressure.

X-ray of the skull showed the previous surgical resection of the mandible, but no evidence of recurrent bone disease. A ventriculogram showed moderately enlarged lateral ventricles. There was localised dilatation and deformity of the left inferior horn with posterior displacement of the 4th ventricle. A bilateral carotid angiogram was normal. A vertebral angiogram showed the basilar artery bowed forward and displaced to the right side. The left posterior cerebral artery was elevated and displaced to the right side, indicating a mass in the region of the cerebellopontine angle (Fig. 1).

Operation. Attempted needle biopsy through the foramen ovale failed, so a craniotomy was undertaken to explore both the middle and posterior fossae. We found a thin vascular tumor, well encapsulated and enclosing a cyst containing dark brown fluid. It extended into the foramen ovale anteriorly and under the tentorium cerebelli posteriorly. Histologically, this was an adamantinoma (Fig. 2).

Postoperative Course. Following this operation the patient's hemiparesis resolved, but the cranial nerve palsies persisted. She remained in good health until March, 1962, when the headaches recurred. At this time she was found to have paralysis of the left 5th, 6th, 7th, 8th and 12th cranial nerves, minimal weakness of the right arm, bilateral papilledema, but no cerebellar signs. Subsequent angiography showed complete occlusion of the left internal carotid artery.

Second Operation. Further craniotomy, while revealing a tense brain, showed no sign of tumor recurrence. Her disabilities continued to increase and she developed in quick succession, paralysis of the left 4th nerve, right homonymous hemianopia, dysarthria and falling vision. Ventriculo-peritoneal anastomosis was performed to reduce the intracranial pressure, but with little improvement in her physical disabilities. Tarsorrhaphy was required to treat a corneal ulcer.

Postoperative Course. In January, 1968, the cerebellar signs recurred and included gross ataxia, nystagmus and incoordination of both arms and legs.

Third Operation. In a desperate attempt to control her increasing disabilities the posterior cranial fossa was reopened. Tumor, partly cystic and partly solid, was seen fixed to the undersurface of the tentorium, extending extradurally into the middle cranial fossa. Partial removal only was possible. Histological examination showed adamantinomatous tissue, exactly similar to the previous sections, with no evidence of malignancy.

Postoperative Course. This operation did not improve her clinical state. She deteriorated slowly and died in December, 1968. Permission for autopsy was unfortunately not obtainable.

Discussion

The adamantinoma or ameloblastoma rarely spreads beyond the peristeme of its original site, unless it is interfered with surgically. It may, however, infiltrate the bone further than is apparent to the naked eye, with the result that excision may be far from adequate, and recurrence only too common. Spread to neighboring organs is unusual, even when the tumor recurs after surgery. Goldwyn et al. reviewed all the cases from the records of 3 of the largest hospitals in Boston, and in 33 patients with histologically proven adamantinomas of the jaw, found 10 new cases and 13 recurrent ones. The patients with recent disease all returned because of a mass or a discharging sinus. A variety of surgical procedures had been attempted. It was evident that only radical surgery produced results suggesting cure;

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radiotherapy was ineffective since the tumor is usually radio-resistant. In spite of many cases of inadequate treatment, there was only one case in this series that showed spread to a neighboring organ. One patient who had received palliative irradiation died 2 years later, and at autopsy the tumor was found to have spread into the middle cranial fossa.

It is doubtful if malignant change ever occurs. Waterworth and Pullar reviewed 7 cases in which lung metastases were reported. These probably occurred by direct spread, from aspiration rather than by lymphatic or vascular dissemination. Small and Waldron, in an extensive review, found that the cases in which malignant change was reported were very poorly documented, especially histologically, and they concluded that the diagnosis of malignancy was extremely doubtful. Schulenberg showed photomicrographs of a case that recurred 3 times over a period of 30 years, yet no changes were seen in the nature of the cells.

Histologically, the adamantinoma is similar to the craniopharyngioma or the basal cell carcinoma that invades the tibia. It is probable that the adamantinoma and the craniopharyngioma are both derived from remnants of the buccal epithelium. The management of intracranial extension of an adamantinoma presents untold difficulties. Involvement of the base of the skull and the issuing cranial nerves makes radical excision impossible, so that one is left to compromise, and thereby achieve what is no more than a palliative operation.