PARATRIGEMINAL EPIDERMOID TUMORS

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Paratrigeminal epidermoid tumors are uncommon but their recognition is important because prompt and proper treatment is associated with excellent results in contrast with many of the other tumors found in this location. We have encountered 2 such tumors. As a survey of the literature has revealed only 1 other, reported by King, 13 a summary of his case is also included. In our first case the lesion was not correctly diagnosed pre-operatively. However, in the second case it was recognized clinically because of its resemblance to the first. In Dr. King’s case a diagnosis of probable meningioma of the middle fossa was made by him and by Dr. Foster Kennedy. It will be seen that with the signs and symptoms plus the radiographic changes a correct pathologic diagnosis may be suspected pre-operatively.

This article deals mainly with epidermoids—so-called cholesteatomas or pearly tumors. To our knowledge the closely related dermoids containing other elements of skin such as sweat glands, sebaceous glands, and hair follicles, have not been found in the area with which we are concerned here. Intracranial epidermoids are relatively rare neoplasms and constitute, according to various figures, between 0.2–1.0 per cent of all intracranial tumors. Mahoney 17 stated that the first intracranial tumor of this type was depicted by Pinson 20 who in 1807 prepared a wax model of such a tumor in the cerebellum and fourth ventricle. Bailey 1 in 1920 collected some 62 cases of which many were doubtful, and Grant and Austin 10 in 1950 reported 22 seen since 1930.

Sex incidence is difficult to determine as in many papers such details are not given, but in those in which the sex of the patients was disclosed, there were 35 males and 26 females. The range in age was from 11–71 years and the average age at the time of surgery as calculated from 39 cases was 35.6 years. The average duration of symptoms was 16 years. In Grant’s 10 series the average duration of symptoms was 2.9 years in the extradural group, and 4.6 years in the intradural epidermoid group.

These are very slowly growing, space-occupying lesions generally agreed to be congenital in origin. 21 They may be epicranial, intradiploic, epidural, intraventricular or intracerebral. The consensus is that they are ectodermal.

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rests, remnants of incomplete separation of neural from surface ectoderm. These fetal inclusions of epidermal cells, depending upon the depth of the layer and/or the embryonic age, result in either dermoids or epidermoids. Closure of the medullary groove in the human embryo occurs at the third week when constrictions appear resulting in the formation of primary cerebral vesicles. During the fourth and fifth week secondary vesicles are developing and through them cell rests may be misplaced away from the midline. The process of formation of the optic and otic vesicles may contribute substantially to the resultant congenital epithelial tumors found in more peripheral locations. It seems likely that the paratrigeminal epidermoids to be discussed here had their origin from embryonic rests associated with the primary otic vesicles.

Intradiploic epidermoids, according to Bucy, are usually found in the frontal and parietal regions. Intracranially epidermoid tumors are found most frequently in the extracerebral spaces along the base of the brain and in the cerebellopontine angle, although Bailey presented 8 cases in which epidermoids were found in the fourth ventricle and 1 in the third ventricle. The symptoms they present are those of any slowly growing tumor. Or they may be found unexpectedly at autopsy. The radiographic appearance is so typical that most intradiploic epidermoids are correctly diagnosed preoperatively. Intracranial extradural tumors may, likewise, present a typical picture of sharply defined bony destruction. In Grant and Austin’s series of 7 extradural epidermoids all showed typical radiographic changes, but of 15 intradural tumors the roentgenograms were of no help in the recognition of 14.

The inner lining of the tumors usually presents a smooth, silky, irregular grey or mother-of-pearl surface. The contents are generally solid, cheesy, dry, and grumous, but occasionally may be a thick, viscid, dark brown or grey material containing cholesterol crystals. In the diploe the epidermoid may have a reddish-pink appearance which King compared to the lining of boiled lobster muscle. The tumors that most closely resemble them are dermoids, as previously mentioned, and the adamantinomas which, however, contain teeth anlage and may contain embryonic teeth that are visible radiologically. Other conditions to be differentiated from the neoplastic epidermoids are the chronic infectious cholesteatomas of the middle ear to which these have no relation but with which they are often confused. Radiologically the intradiploic lesions may also be confused with sarcomas of the skull, eosinophilic granulomas, and osteitis fibrosa cystica.

Microscopic examination of these tumors reveals a flat stratified squamous epithelium resting on a supporting connective-tissue membrane without papillae. There are no dermal elements, such as sweat and sebaceous glands and hair follicles. The surface of the epithelium is turned inward so that as the surface cells are continuously cast off they form the cheesy interior of the tumor which gradually increases in size.

Theoretically the ideal treatment of any epidermoid is complete removal.