ECCHORDOSIS PHYSALIPHORA SPHENO- OCCIPITALIS.

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THE occurrence of small, transparent, jelly-like nodules projecting into the interior of the skull from the middle of the clivus blumenbachii (dorsum sellæ) was first described by Luschka² in 1856. In the following year Virchow⁸ gave a detailed and illustrated account of the condition to which he applied the term "Ecchondrosis physalifora," in the belief that the protrusions were of cartilaginous origin and nature. Müller³ (1858) first suggested a notochordal origin, and adduced important embryological and anatomical evidence in support of this view, but it was not until nearly forty years later that Ribbert⁴ (1894),⁵ (1895), brought forward the final proof of the theory. In addition to studying a series of five cases of spheno-occipital ecchondrosis (as it was still called), and drawing important conclusions therefrom, he successfully reproduced the condition in rabbits by puncture of the intervertebral discs. This was followed by hernia of the nucleus pulposus, and the nodule of tissue so formed showed evidence of cellular proliferation after a time, and a definite increase in size. After a year it had become largely absorbed, but its histological structure accorded so closely with that of the spheno-occipital protrusions already described that there could be no doubt as to the identity of nature and origin of the two formations. Ribbert proposed the term 'chordoma.'

Except for the case of Klebs, where the tumour was the direct cause of death, all the examples of 'ecchondrosis' so far reported had been casual findings in the post-mortem room. The notochordal nodules were of such small size and soft consistence as not to give rise to pressure symptoms, still less to a fatal issue. From 1903 onwards, however, reported cases of chordoma have nearly all been of frankly neoplastic character and of clinical importance, the tumours being of large size and causing serious pressure effects.

From the clinical standpoint it seems desirable that a clear distinction should be drawn between these two conditions—the small jelly-like nodules with very limited powers of growth, and the large progressive formations which give rise to symptoms and in time cause death. The former are of the nature of notochordal protrusions rather than tumours; the latter are genuine neoplasms, possessing in most cases many of the

FIG. 1.—‘*Echordosis physaliphora speno-occipitalis*’; a protrusion and limited proliferation of notochordal tissue through the middle of the clivus. There is an aperture in the dura mater at this point.

FIG. 2.—Pons, medulla and cerebellum from the same case as *Fig. 1*. Part of the notochordal protrusion is adherent to the basilar artery, and has been torn away from the rest in the removal of the brain.

stigmata of malignancy. We would suggest that the term 'chordoma' be reserved for the second group, and that a variant of Virchow's term, 'Ecchordosis physaliphora,' might be applied to the others. It may be that there are intermediate grades, or that chordoma proper arises in and from a pre-existing ecchordosis, but the important practical point is that the clean-cut division suggested is entirely justified by all the cases of both types so far recorded.

Ribbert ⁶ (1904) has stated that ecchordosis sphenoccipitalis is a thing of common occurrence, and claimed to have found it himself in 2 per cent. of autopsies. So far as we are aware this high incidence rate has not been confirmed by others, and as the matter is of considerable theoretical importance we thought it worth while to try to obtain further evidence. In a series of 200 specially investigated autopsies, we have found three examples of sphenoccipital ecchordosis, all of them of small size, an incidence rate of 1.5 per cent. In each, the little gelatinous nodule arose from the middle line of the clivus, about half an inch behind the posterior margin of the pituitary fossa, and projected through an aperture in the dura mater. In each, also, there was adhesion to the basilar artery, so that in two cases the nodule was torn across in removing the brain from the skull (*Figs. 1 and 2*), while in the third it separated from the bone and adhered to the artery in its entirety.

The first (and largest) specimen (*Figs. 1 and 2*) was from a woman of fifty-nine, who died of advanced polycystic disease of the kidneys and liver; the second, which was much smaller, from a man of seventy-one, who died of pyæmia following suicidal cut-throat. He was suffering at the time from advanced silicosis and phthisis. The third (*Fig. 3*) was also very small in size, and was from a woman of sixty, who died of cancer of the pharynx. *Fig. 4* shows the tiny aperture in the dura mater

FIG. 3.—A small, bilobed ecchordosis physaliphora is seen adherent to the basilar artery.

of the dorsum sellæ present in this case. The ecchordosis, which consisted of two tiny jelly-like nodules adherent to the basilar artery, lay immediately over the aperture, through which it had presumably made its way from the subjacent bone.

In a fourth case, where no extra-osseous protrusion of notochordal tissue had occurred, a small projecting bony nodule in the middle line of the clivus was found to contain in its interior a little mass of similar clear, transparent jelly-like tissue.

FIG. 4.—The clivus blumenbachii (dorsum sellæ) of the same case, showing a tiny aperture in the dura mater exactly opposite the position of the ecchordosis.

Microscopically, all these nodules consist of a number of highly vacuolated, mucin-containing cells, showing little or no evidence of active growth, with, here and there, large intercellular collections of homogeneous secretion, only some of which gives the staining reactions of mucin. In sections stained with hæmatoxylin and eosin the appearance of the more cellular areas is not unlike that of

adipose tissue, and is almost exactly like the section of chick-embryo notochord figured in the last edition of Stöhr's *Histologie*.⁷

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