EYE SIGNS IN PINEAL TUMORS

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Tumors of the pineal body are rare. According to Cushing's figures, 0.7 per cent of his 2,000 intracranial tumors were pineal in origin. In spite of their rarity, however, certain clinical manifestations of growths in this region are of more than usual interest. Among these features are the disturbances concerned with pupillary activity, movements of the eyeballs and other ocular disorders. The paper here presented is devoted chiefly to a clinical study of these oculomotor and other related disturbances.

The material from which this information has been gathered consisted of 16 cases in which a diagnosis of pineal tumor was made at the Lahey Clinic during the years 1933 to 1944 inclusive. In 11 of the 16 patients the lesion was verified histologically either from tissue removed at operation or at autopsy. In the other 5 patients a presumed and almost certain diagnosis was made on the basis of the clinical evidence together with the finding of the characteristic filling defect in the posterior portion of the third ventricle as seen in the ventriculogram. These diagnostic features were supported by the demonstration of internal hydrocephalus and the complete relief of symptoms and signs following a decompression and roentgen therapy. In spite of the fact that it was not possible to verify the tumor histologically in these 5 patients, we feel that the clinical picture, the ventriculogram and the course following decompression and radiation therapy all support the diagnosis of pineal tumor with sufficient certainty to include them in this survey.

The surgical removal of a pineal tumor is an extremely difficult task and entails considerable risk. On the other hand, as Horrax and Daniels have pointed out, subtemporal decompression followed by radiation therapy offers a comparatively safe method of treatment which has proved highly satisfactory in several cases. For this reason it is highly important to obtain all available evidence bearing upon the diagnosis of a growth in the pineal region. The oculomotor and pupillary disturbances furnish considerable information along these lines.

Before taking up in detail the various ocular disorders that may occur with pineal tumors, it may be well to note briefly certain general features shown by patients harboring these lesions. By recalling the strategic position of the epiphysis just above the corpora quadrigemina (Fig. 1), one can predict with a good deal of accuracy the most frequent signs and symptoms resulting from a tumor of this structure:

1. Increased intracranial pressure, often acute, producing headache,
nausea, vomiting and papilledema. Mechanism: compression and obstruction of the aqueduct of Sylvius with resulting internal hydrocephalus.

2. Pressure on the corpora quadrigemina, giving rise to:
   (a) Eye signs.
       (1) Impaired pupillary reactions.
       (2) Limitations of extra-ocular movements, especially conjugate movements upward.
       (3) Nystagmus.
       (4) Strabismus.
   (b) Central deafness. This is now believed to be due not entirely to pressure on the inferior colliculi but also to involvement of the lateral lemnisci.

3. Cerebellar signs, by transmitted pressure downward against the tentorium.

   In addition to the foregoing features, certain other general disturbances of function may be present. These are:

   1. Possible endocrine dysfunction in the form of macrogenitosomia praecox. This is a relatively infrequent condition and, when present, has been observed only in males under the age of puberty. Of the 6 patients under the age of fifteen in this series, only one (C.C.) showed this syndrome. In Russell and Sachs' review of the literature there were 17 patients under fifteen years of age with the diagnosis of a pineal tumor. Of this number, 3 presented the clinical picture of macrogenitosomia praecox.