TIC DOULOUREUX AND ITS RELATIONSHIP TO TUMORS OF THE POSTERIOR FOSSA
ANALYSIS OF TWENTY-FOUR CASES

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Since the introduction by Dandy\(^1\) of the cerebellar approach for section of the sensory root of the trigeminal nerve, the advantage of this route in the disclosure of organic lesions that might account for the production of tic douloureux has been pointed out several times.\(^2,3\) Dandy\(^4\) found that 60 per cent of his patients operated upon by the cerebellar approach showed definite abnormalities such as tumors, aneurysms, angiomas, aberrant vessels on the nerve, congenital anomalies, and adhesions between the sensory root and brain stem. For detailed figures regarding these abnormalities the reader is referred to his paper.

It is the purpose of this communication to analyze only that group of cases in which tumors were found incidentally upon sectioning the sensory root at the pons for tic douloureux.

From 1925 to 1945 there were 757 cases diagnosed as tic douloureux at the Johns Hopkins Hospital, and in 473 of these, operation was performed by the cerebellar approach. In the whole series 58 per cent were females and 42 per cent were males. The ages ranged from 30 to 70 years. In 24 (5.1 per cent), tumor was found in the neighborhood of the 5th nerve. Three types of tumors were encountered: neurinomas, epidermoid cysts and meningiomas.

GROUP I. NEURINOMAS

Eleven neurinomas (46 per cent) were found in this series. These presumably arose from the 8th nerve. Males and females were about equally affected. Eight of the 11 patients were between the ages of 50 and 70. The chief complaint on admission in all cases was intermittent, lancinating pain in the distribution of one or more branches of the trigeminal nerve. The 3rd branch alone was not affected in any case, the 1st branch in 1, the 2nd branch was involved in 3, the 1st and 2nd in 1, the 2nd and 3rd in 6, and all branches in 1. There was no significant predominance of sides, 6 occurring on the right and 5 on the left. The pain was typical of tic douloureux with definite trigger zones and was precipitated by swallowing, chewing, washing of the face, etc. Ten patients showed associated loss of hearing on the same side and in the remaining case there was no impairment: the hearing deficiency was not found significant in this age group. Loss of hearing preceded pain in 8 cases by an average of 7.5 years. In 1 case tic was present 11 years before impairment of hearing and in another both the 5th and 8th nerves were affected simultaneously. Tinnitus on the affected side was present in
only 5 cases and bilateral tinnitus in 1. In 1 case the tinnitus, which had been present for 7 years, abruptly disappeared with the onset of pain. Dizziness was present in 1 case. Dysphagia occurred only once in this series, this being in the one case that did not show any impairment of hearing. There was a history of gustatory fits in the affected side in 1 case. In 10 cases the neurologic examination corroborated the subjective impairment of hearing, this being moderate for low tones and quite pronounced for high tones. Vestibular tests were performed in 6 of the 10 patients: 5 had loss of vestibular response on the same side and 1 on both sides. Mild sensory changes in the affected side of the face were found in 4. In 1 case there was bilateral papilledema with unilateral deafness but there were no sensory changes.

The diagnosis of tumor of the cerebellopontine angle was suspected in 6 cases and accordingly X-rays of the skull were taken. Erosion of the porus acusticus was definite in 3.

The unilateral cerebellar approach as originally described by Dandy was used in each case. The tumor was totally removed in all cases. It was usually small, the average weight being 6.28 gm., the largest 15.6 gm. and the smallest 0.1 gm. The gross and microscopic appearance of the tumors was that of typical neurinoma. All these tumors, with the possible exception of one, seemed to arise from the porus acusticus and pressed on the 5th nerve in varying degrees. The 5th nerve was sectioned in all but 2 and in these 2 there was complete relief of pain after the operation. There was 1 fatality in the series. Death occurred a few hours after the operation, and autopsy revealed edema of the medulla and pons with small hemorrhages. The rest of the patients showed complete relief of symptoms after operation.

Comment. Eleven cases of neurinoma producing typical tic douloureux have been summarized. The chief complaint on admission in all these cases was that of pain in one side of the face. In 10 cases hearing was impaired on the same side of the lesion, which in an age group of from 50 to 70 years is apt to be minimized. Mild sensory changes were found in 4, and in 1 there was bilateral papilledema with unilateral deafness but without sensory changes. X-rays of the skull were taken in 6, only 3 showing definite erosion of the porus acusticus. Most of these cases could easily have been diagnosed as "idiopathic" tic douloureux and the nature of the underlying condition could have been missed had it not been for the route of approach selected. A vivid example is Case 4, summarized below, in which section of the 5th nerve was performed by the subtemporal route in another clinic, with recurrence of pain 2 years later. At operation through the cerebellar approach a neurinoma was found. It can be safely concluded that all cases of tic douloureux showing significant impairment of hearing on the same side should be considered potentially as neurinomas of the cerebellopontine angle possibly arising from the porus acusticus.

Case 1. #15400. Mr. W. H., a white male aged 67, was admitted July 27, 1927 with the chief complaint of pain for 2 years in the right side of the face, starting in the right temple with radiation to the lower jaw. The pain was lancinating and intermittent with trigger zones in
the cheek, and was precipitated by washing the face, swallowing and chewing. Alcohol injections given on 4 occasions relieved his pain temporarily. For 12 years there had been loss of hearing on the right.

Neurologic examination on admission was negative except for moderate diminution of hearing on the right. The preoperative diagnosis was tic douloureux.

Operation: Unilateral cerebellar approach, right, at which a small tumor occupying the upper half of the fossa, mainly above the 8th nerve, was found. The 5th nerve was pushed upward and it was sectioned. Weight of tumor: 5 gm.

Histologic diagnosis: Neurinoma.

Case 3. #U-33213. Mrs. C. S., a white female aged 50, was admitted Sept. 22, 1930 with the chief complaint of pain in the left side of the face, intermittent, lancinating in character, and of 2 years' duration. Pain was localized to the cheek and upper gum on the left. In addition there had been gradual diminution of hearing on the left with constant tinnitus, described as roaring, for 10 years.

Neurologic examination showed bilateral papilledema of 2-3 diopters. There was total left deafness with diminished caloric response on this same side. Deep reflexes were hyperactive bilaterally. X-ray of the skull was negative.

Operation: Unilateral cerebellar approach, left. A small deep-seated tumor was found at the angle, apparently arising from the 8th nerve, extending medially beneath the brain stem. The tumor was completely enucleated and the sensory root of the 5th sectioned. Weight of tumor not reported.

Histologic diagnosis: Neurinoma.

Case 4. #U-44319. Mr. J. A., a white male aged 60, was admitted July 11, 1932 with the chief complaint of tic douloureux on the right, affecting the 3 branches equally with trigger zones in the 1st, for 18 years. Six years previously section of the 5th nerve on the right by the subtemporal approach had been done elsewhere with relief of symptoms for 2 years, after which pain recurred affecting only the 1st branch on the same side. He also complained of an intermittent bad taste on the right side of the tongue. Twenty years previous to admission he had punctured his right ear drum in attempting to remove a piece of coal, and following this there was progressive deafness on the right.

Neurologic examination show hypalgesia and hypaesthesia in the 3 branches of the right 5th, obviously from previous operative procedure, complete right deafness, hyperactive deep reflexes on the left, and no response to caloric vestibular stimulation on either side. X-ray of the skull was negative. Preoperative diagnosis: recurrent tic douloureux.

Operation: Unilateral cerebellar approach, right. A hard tumor was found at the angle, apparently arising from the 8th nerve, and was completely enucleated. Weight: 15.6 gm.

Histologic diagnosis: Neurinoma.

GROUP II. EPIDERMOID CYSTS (CHOLESTEATOMAS OR PEARLY BODY TUMORS)

Nine (38 per cent) such tumors were found in our series of 24. Males and females were equally affected. Their ages were between 30 and 60. The chief complaint on admission was that of intermittent, fleeting, lancinating pain in one side of the face, the left side being predominantly affected, only 1 case occurring in the right. Not a single branch was affected selectively; all 3 branches were involved in 7, and the 2nd and 3rd in 2. In 1 case the pain alternated at times from one branch to the other. Remissions of attacks were rare, occurring in only 3 cases and varying from 3 to 7 years. The average time since onset of symptoms was 8 years, the longest 23 years and the
shortest 6 weeks. There was no impairment of hearing in any instance. One case gave a history of intermittent clonic contraction of the affected side of the face. There were no headaches. The neurologic examination was negative in practically every case except for slight facial weakness in 1, horizontal nystagmus with diminished caloric vestibular response in another, and loss of taste on the anterior two thirds of the tongue in another. X-rays of the skull were taken in 2 cases in which the diagnosis of a tumor at the angle was suspected, but these were reported as normal.

The unilateral cerebellar approach was performed in all cases. The tumors presented readily at the angle and all of them but 2 were described as large growths extending from the tentorium in front to the foramen magnum behind and beneath the brain stem medially. The 5th nerve ran through the tumor in all cases and there was involvement of the 7th, 8th, 9th and 10th in all but 2. All were typical pearly body tumors and in their extirpation the contents were aspirated by suction, only the capsule being saved for histologic sections, so that a fair estimate of the weight of these tumors is not available. A complete removal was obtained in every case and section of the sensory branch of the 5th nerve performed in all. There were no mortalities. Every patient was completely relieved after the operation.

Comment. In contradistinction to Group I there was no impairment of hearing in any of these cases. The left side was almost selectively affected and the pain involved the 3 branches of the 5th nerve in every case except 1. The correct diagnosis was suspected in 2 but no changes were demonstrated by X-rays. The true nature of the lesion could have been completely missed had the cerebellar approach not been used. This stresses even more strongly the importance of this route in cases of tic douloureux, as emphasized so often by Dr. Dandy.

Case 14. #U-29928. Mr. J. A., a colored male aged 35, was admitted Mar. 8, 1930 with the chief complaint of paroxysmal attacks of sharp shooting pain in the left side of the face for 4 years. The pain affected all 3 branches but chiefly the 2nd and 3rd. Rubbing the face would precipitate an attack. Recently the attacks had been more frequent, 10 to 12 a day.

Neurologic examination was negative. Hearing and vestibular function were normal with no sensory changes over the face. Preoperative diagnosis: tic douloureux.

Operation: Unilateral cerebellar approach, left. A typical pearly body tumor was found at the angle. It pushed the 5th nerve downward, bending it markedly, and did not project below the 8th. The 5th nerve ran directly into the tumor. The sensory branch was divided. The tumor extended medially to the midbrain, going beneath it for some distance. The contents were suctioned away and the capsule weighed 2½ gm.

Histologic diagnosis: Epidermoid cyst.

Case 18. #123347. Mrs. M. S., a white female aged 51, was admitted on Oct. 20, 1937 with the chief complaint of paroxysmal lancinating pain in the right side of the face for 8 years. The pain was intermittent and typical of tic douloureux. The 3rd branch was primarily affected but later also the 2nd. In addition to these attacks of pain there was intermittent clonic contraction of the right side of the face, occurring independently of the pain and lasting for a few seconds.

Neurologic examination showed twitching of the right side of the face with closure of the eyelids in clonic contractions, right facial weakness and a trigger zone in the cheek. There
was no loss of hearing except for diminution of the threshold for high tones bilaterally. Vestibular response was absent on the right. X-rays were not taken.

Operation: Unilateral cerebellar approach, right, disclosing a large pearly body tumor at the angle. The 5th nerve ran directly through it. It extended medially beneath the brain stem. The 8th nerve ran directly across the tumor. The tumor was removed piecemeal and part of it was suctioned away. Weight of the part saved was 6.9 gm.

Histologic diagnosis: Epidermoid cyst.

GROUP III. MENINGIOMAS

In this group there were 4 cases (16 per cent of the series). They were equally divided between males and females, the ages ranging between 47 and 71 years. There was no predominance as to the side affected. The average duration of symptoms was 5.5 years, the shortest being 9 months and the longest 8 years. Symptomatology was characterized by typical intermittent episodes of pain of lancinating character localized in one side of the face and involving the 2nd and 3rd branches in 2 cases and all branches in the remaining 2. As in cases of tic douloureux, pain was precipitated by washing the face, chewing, swallowing and talking, with definite trigger zones. There was bilateral loss of hearing in 1 case with impairment of vestibular function on the side of the lesion. Hearing was not impaired in the remaining cases nor was there involvement of any other cranial nerve. History of headache was elicited in 1.

The usual unilateral cerebellar approach was used in each case and small encapsulated tumors were found pushing the sensory root posteriorly, in all probabilities arising from the tentorium. The tumor was totally enucleated in 2 cases, partially in 1 and was not extirpated in the other. Specimens were lost in 2 cases but their gross appearance and attachment suggested a meningioma. The other 2 were typical fibroblastic meningiomas. Weight of the tumors was not recorded. The sensory root of the 5th nerve was totally divided in all cases. One patient in whom the tumor was partially removed died 3 days postoperatively. During the operative procedure it was necessary to cauterize the posterior inferior cerebellar artery and the patient never regained consciousness. At autopsy tumor was seen projecting from the posterior aspect of the body of the sphenoid bone, extending to the left along the posterior surface of the petrous bone posteriorly, and laterally to the right of the clivus. The base of the tumor was attached to the dura, where there was a thick layer of newly formed bone. There was softening of the left half of the medulla and pons. The remaining 3 patients had complete relief of symptoms after operation.

Comment. Four cases of meningioma at the cerebellopontine angle producing tic douloureux have been summarized. The primary symptom was that of intermittent attacks of lancinating pain in one side of the face. There was impairment of hearing in 1, the other 3 showing no evidence of other cranial nerve involvement. The preoperative diagnosis in all was that of tic douloureux as there was nothing in the clinical history of the patients to suggest a tumor of the posterior fossa.
Case 22. #U-42534. Mr. N. B., a white male aged 67, was admitted April 1, 1932 with the chief complaint of periodic paroxysmal pain in the right side of the face of 2 years' duration. The pain involved chiefly the areas of distribution of the 2nd and 3rd, less frequently the 1st. It was precipitated by talking, laughing, or rubbing his face.

Neurologic examination was essentially normal. Preoperative diagnosis: tic douloureux.

Operation: Unilateral cerebellar approach, left. A small tumor was disclosed filling the angle between the tentorium and the 8th nerve and attached to the dura. The 5th nerve was pushed posteriorly and made almost a semi-circle at its entrance to the pons. The sensory root was divided. The tumor was totally extirpated. Weight not stated.

Histologic diagnosis: Fibroblastic meningioma.

DISCUSSION

From the preceding analysis of cases it may be clearly seen that the relationship between tic douloureux and tumors of the posterior fossa is more than casual and for one reason or another the 5th nerve has been selectively affected. In the neurinomas, the diagnosis could have been suspected if the hearing changes had been properly evaluated, but they were taken lightly due to the frequency of disturbances in hearing in otherwise normal elderly individuals. In the epidermoid cysts, however, there was nothing to suggest a tumor with the possible exception of 1 case, and one wonders why the 5th nerve alone gave irritative symptoms with sparing of the 7th, 8th, 9th and 10th, which were found to be similarly involved at operation. Had these patients been operated upon by the subtemporal approach, the underlying pathology would have been missed, as did happen in 1 of the cases. This aspect alone, therefore, would recommend the cerebellar approach over any other procedure. But the question then arises as to how many tumors of the Gasserian ganglion would be missed if the posterior route were exclusively employed. These tumors are relatively rare in comparison to tumors of the posterior fossa and in contradistinction they give unmistakable neurologic signs of their presence. Apprehension has been postulated by others regarding the use of the posterior approach due to technical difficulties, but in skilled hands there is nothing to fear. The change from the prone to the lateral position in the unilateral approach as suggested recently by Mount and used by Otenasek has made this a relatively easy and rapid procedure. There are, furthermore, other advantages of this approach as enumerated by Dandy which are not within the scope of this paper, but from the evidence presented it can be clearly seen that many pathological lesions involving the sensory root which would otherwise be missed will be disclosed by this approach.

SUMMARY

1. A total of 24 tumors of the posterior fossa producing tic douloureux have been reported, 46 per cent of which were neurinomas, 38 per cent epidermoid cysts, and 16 per cent meningiomas.

2. Each group of tumors has been analyzed.

3. All of these tumors would have been missed if the subtemporal route had been employed.
4. The merit of the cerebellar approach as originated by Dandy for section of the sensory root of the 5th nerve has been reemphasized.

REFERENCES

6. OTENASEK, F. J. Unpublished data.