Clinical features of the epidermoids of the basal cisterns of the brain¹

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Few intracranial tumours have such a great clinical interest as the so-called pearly tumours of Cruveilhier. Described by this French pathologist under this name, in 1829, many reports have since appeared in the medical literature.

The history of our knowledge in this field is well known and only a few comments will be made. A certain confusion started when Müller (1838) proposed the term 'cholesteatoma' to describe a tumour of the cranial bone containing masses of cholesterol inside. Later, many authors found some apparently similar formations related to chronic infections, mainly from the mastoid and middle ear. Further, Virchow proposed, in 1855, a metaplastic endothelial origin for these lesions.

However, Bostroem, in 1897, clarified the situation with his study of 32 observations from the literature and some personal ones. There are typical tumours formed from misplaced epithelial rests during the first weeks of the embryonic growth and these may be either epidermoids or dermoids. In the first group, the capsule is formed only by epithelial layers and these are much more frequently found than the dermoids containing other dermal elements, such as hair, sweat glands, etc. Percival Bailey, in 1920, studied histologically the different elements of the capsule of the epidermoids and since then many other papers have appeared in the literature.

On the other hand the term 'cholesteatomas' has been restricted to those inflammatory lesions appearing mainly in the mastoid region and in relation to chronic middle ear infections. There are, however, some rare epidermoids localized in the petrous bone, the origin of which is still not completely known. Some seem to be real inclusion epidermoid tumours but others appear in patients with chronic ear infections, as has been remarked by Tytus and Pennybacker (1956) in the consideration of six personal cases.

Epidermoids have attracted the attention of the neurosurgeons for many years. Dandy even called

them the most beautiful tumours of the body. Here we have strikingly large tumours with a white and glistening appearance resembling mother of pearl. The inside of the tumour is formed by masses of epithelial debris and desquamating keratin and cells derived from the epithelial capsule—'ghosts of epithelial cells', to use Bailey's expression—which very slowly accumulate inside of the capsule and mix with fatty material.

Because of these circumstances the growth of these tumours is very slow and thus may reach a considerable size before producing clinical symptoms. In fact the 'princeps' observation of Cruveilhier was an incidental necropsy finding in a man who died of a head injury. As can be seen in the necropsies the epidermoids adapt themselves to all the available space inside the cranium and spread widely along the brain structures. In the anatomopathological study of seven cases, studied at necropsy in the National Hospital, Queen Square (Ulrich, 1964), these points were emphasized and also that these tumours tend to grow along a plane of cleavage between the nerve fibres. Thus, elongated finger-like intracerebral extensions of the epidermoid may sometimes be responsible for the first symptoms, such as third nerve palsy and hemiparesis in two of the cases of Ulrich (1964).

The clinical literature of this kind of tumour is very large indeed and can be found in different reviews (Mahoney, 1936; Tytus and Pennybacker, 1956; Lepoivre and Pertuiset, 1957; Tönnis, 1962; Olivecrona, 1967; etc.).

The relative frequency of the epidermoids in relation to other intracranial tumours is between 0.5 to 1.8% of the neurosurgical series.

In our personal material of 3,420 brain tumours and other space-occupying lesions, excluding vascular lesions, that have been operated on in our neurosurgical services of Madrid from October 1946 to December 1968 we have a total of 60 epidermoids that may be classified into the provisional groups shown in Table I according to the clinical and surgical data.

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TABLE I
CLASSIFICATION OF 60 EPIDERMIDS

Localization of 60 epidermoids or pearly tumours	Cranial bones 15 (25%)			
	Intradural 45	Base of the brain 40	Suprasellar- chiasmatic	9
			Parasellar-Sylvian	9
			Retrosellar cerebello-pontine	19
			Basilar-posterior fossa	3
	Ventricular system 5		Lateral and III ventricles	3
			IV ventricle	2

We have adopted this simplified classification because it is our belief that it is not possible to make a strict topographical separation of the epidermoids. The majority of the neurosurgeons have spoken of cerebello-pontine, suprasellar, temporal, cerebellar, and many other sites when referring to the sub- or intra-dural epidermoids. But this separation reflects only the most obvious symptomatology or the surgical approach used to reach part of the tumour. But we know from our surgical experience and the necropsy material that these tumours extend so widely across the brain that their real localization cannot be obtained either from the clinical symptoms and signs or even from the data obtained at the surgical procedure.

Lepoivre and Pertuiset (1957) have adopted a more rational classification of the epidermoids into three main groups and have correlated them with the main blood vessels at the base (vertebro-basilar, carotid) and the choroid arteries. The result of this classification is as follows: retrosellar or vertebro-basilar; suprasellar, parasellar, or carotid; intra-ventricular or choroid.

This classification of Lepoivre and Pertuiset (1957) is a great advance and there is no doubt about the separation of the intraventricular group that represents between 10 to 20% of the intradural epidermoids of the different neurosurgical series.

However, the separation of the epidermoids located in the basal cisterns is more difficult, and personally we believe that it would be better to unify all these basilar localizations and then try to explain the irregular and multiform symptomatology of these curious slowly growing tumours.

The classical pathologists, like Cruveilhier, Bostroem, and others, had already observed that the pearly tumours appeared mainly at the base of the brain and this fact has been confirmed in later

years. Lepoivre and Pertuiset (1957) found a figure of 75% and Tönnis (1962) one of 80% for such a localization. Other pathologists have recently insisted on the more frequent localization of the epidermoids in the sub-arachnoid cisterns somewhere near the midline of the base (Rosenbluth and Lichtenstein, 1960).

SYMPTOMS AND SIGNS

Taking as a whole group our 40 patients with epidermoids located at the sub-arachnoid cisterns of the base, we see the great variety and diversity of the main symptoms and signs recorded in the clinical histories.

The sex of the patients showed no significant difference, as we had 19 males and 21 females. An analysis of age revealed the following: under 20 years, seven patients; between 20 to 40 years, 20 patients; and between 40 to 64, 13 patients.

The duration of the clinical histories was as follows: less than one year, nine patients; from one to five years, 16 patients; from six to 10 years, eight patients; from 12 to 25 years, seven patients.

These data show that these tumours, in spite of their congenital origin, may start to give symptoms at practically any age and may remain asymptomatic for long periods of the life of the patients. However, taking into consideration the ages of the patients when they come to the neurosurgeons and the long histories present in many of them, we may conclude that in the majority of patients symptoms first appear at an early age. Moreover, long histories extending for periods of several years appeared in nearly 75% of the cases and in nearly 20% for periods of 10 to even 25 years.

These two general facts—the clinical presentation at any age but most frequently between 20 to 40 years, and the long histories of many years' duration in a fair number of the patients—have been emphasized in many papers, cf. Ulrich (1964).

The presence of papilloedema was recorded in only 15 of our patients. This figure shows again that many of these basal tumours may attain a great size without inducing signs of increased intracranial pressure because of their typical slow growth through the accumulation of desquamating cells and keratin inside the capsule.

Because of the situation and extension of the epidermoids through the basal cisterns the clinical signs are very diverse and bizarre. We can make a list (Table II) of the main ones recorded in the histories of the patients.

This long list demonstrates the possible involvement of many different nervous structures from the optic chiasma to the medulla (Fig. 1). However, the trigeminal and facial nerves are more often affected

FIG. 1. Possible involvement of different nervous structures in the epidermoids of the basal cisterns.

TABLE II
CLINICAL SIGNS OF EPIDERMIDS

Primary optic atrophy and hemianopias	6
Epileptic attacks (generalized or focal, uncinat)	8
Hemiparesis	8
Speech disturbances	3
Diplopia and oculomotor disturbances	5
Trigeminal nerve involvement	17
Trigeminal neuralgia	7
Facial nerve involvement	17
	4 hemifacial spasms)
VIII nerve involvement	12
IX, X and XI nerves involvement	5
XII nerve involvement	2
Nystagmus	13
Cerebellar disturbances	13
Dysarthria	3
Long motor tracts involvement with bilateral Babinski sign	6

and sometimes the irritation of one of these nerves (trigeminal neuralgia or hemifacial spasms) or their involvement precedes for many years the rest of the symptoms and signs. Also auditory-vestibular symptomatology, together with nystagmus and cerebellar signs, are very prominent in these epidermoids of the base of the brain.

An interesting finding is that the protein content of the cerebrospinal fluid is usually normal, as remarked by Tytus and Pennybacker (1956) and others in the cerebello-pontine syndromes produced by these tumours. The cerebrospinal fluid was studied in six of our cases and the amount of

protein was increased in only one of them with a main parasellar-Sylvian localization.

CLINICAL SYNDROMES

We have seen the great number of symptoms and signs that appear in these tumours and the difficulty of establishing clear-cut syndromes, for the symptoms and signs may reveal only that some nervous structures have been affected—usually some of the cranial nerves—but not the real extension of the tumour throughout the basal cisterns. We may perhaps attempt to separate four main groups according to the predominant symptomatology; this can also be related to whether the growth extends more anteriorly, laterally, or posteriorly.

Those extending forward produce clinical syndromes of the chiasmatic region with loss of vision, optic atrophy, and temporal hemianopias. Olivecrona (1932) and others have described this clinical syndrome of the suprasellar epidermoids. The differential diagnosis with other tumours of this region has also been thoroughly discussed in the literature.

Sometimes the anterior extension of the epidermoids of the base is more lateral and produces a clinical syndrome of the temporal or Sylvian region (parasellar epidermoids). Usually there are in those cases a history of epilepsy with generalized or focal

FIG. 2. Clinical signs of 16 cases with clear tumoural syndromes of the cerebello-pontine angle.

attacks (uncinate or Jacksonian seizures) accompanied by hemiparesis and, more rarely, by speech disturbances. Involvement of the Vth nerve was observed in about one-third of these cases and more rarely of the oculomotor nerves. One patient presented with what was an apparently typical trigeminal neuralgia. This was successfully treated by subtemporal retrogasserian section of the sensory root and was followed by an unspecific meningeal infection a long time before other signs of oculomotor involvement led to the right diagnosis.

Even more frequently the epidermoids of the base produce clinical syndromes of the cerebello-pontine angle. In fact, reviewing the different neurosurgical series of the literature, we found that a clear clinical syndrome of this region appears in about 20 to 40% of the total number of epidermoids (more than 40% of our material of intradural localizations).

In some cases we saw only the clinical picture of a trigeminal neuralgia, and it is well known by neurosurgeons that, approaching the sensory root by the posterior sub-occipital route, we sometimes find an epidermoid pressing on the trigeminal nerve. Dandy (1945), Olivecrona (1949), and many others have remarked on this fact. In our material, we have altogether seven patients (one already mentioned) in whom the clinical picture started as a

trigeminal neuralgia, and five of them were treated as such before other symptoms appeared.

The rest of the cerebello-pontine angle symptomatology was very obvious in 16 patients of our material of epidermoids. The most frequent initial symptoms were related to the VIIIth nerve (hypacusia, ear noises) in half of the cases. An interesting initial symptom corresponded with the facial nerve in four patients, three of them with different degrees of facial palsies and one with hemifacial spasm. We have mentioned that, for a considerable time, trigeminal neuralgia may be the only symptom of the epidermoids and may be followed later by other tumoural signs of the cerebello-pontine angle (three cases). Other initial symptoms (headache, difficulty in swallowing, etc.) appear only occasionally.

The clinical signs of our 16 cases with clear tumoural syndromes of the cerebello-pontine angle have been analysed in another paper (Obrador, Boixadós, Queimadelos, Córdoba, and López-Zafra, 1968) and may be summarized as in Figure 2.

All the clinico-topographical localizations that have been outlined are very schematic and do not in any way represent the true extension of the tumour. It has been repeatedly mentioned how the slow growth of these tumours by the accumulation of desquamating epithelial cells allows their accom-

modation and adaptation within the skull and in relation to the nervous structures. As many other neurosurgeons, we too have been surprised to see at operation the enormous extension of these basal epidermoids. Some of them with a unilateral symptomatology of the cerebello-pontine angle extend through the tentorium into the supratentorial region in front of the pons. Others inducing a more anterior supra or parasellar symptomatology may also have very wide posterior retrosellar extensions.

We have, in fact, three examples of what may be considered as the most extensive basilar-posterior fossa epidermoids. In these patients there was clear dysarthria, nystagmus, cerebellar signs, and bilateral pyramidal signs with plantar extensor responses. Diplopia, hemiparesis, a profound spasmodic laughter, and some involvement of the nerves of the angle on one side were also recorded in one patient. It is interesting that a wrong diagnosis of multiple sclerosis was made in two of these patients—especially in one of them with normal optic discs.

Two of these cases were studied at necropsy and the epidermoid extended very widely through the base of the brain around the midbrain, pons, and medulla and growing forward to the sellar and parasellar regions. In the other patient, the tumour was surgically approached through a sub-occipital craniotomy and filled the cisterna magna and both cerebello-pontine angles.

In the seven post-mortem observations of Ulrich (1964), the tumours could also be included in the so-called retrosellar variety but usually they extended forward and were not confined to the posterior fossa.

Neuroradiological studies—and especially the visualization of the cisterns with air and the use of vertebral angiography—may be very useful in demonstrating the real extension of the tumour at the base of the brain, as we have seen in our material.

SUMMARY

A study of 40 epidermoids of the base of the brain verified at operation or necropsy demonstrates that the clinical manifestations often do not indicate the wide extent and site of the tumour but merely

some damage or irritation of some nervous structures, especially the trigeminal, facial and auditory nerves, the cerebellum, and optic pathways. Epileptiform phenomena, motor tract involvement at different levels, speech disturbances, and other cranial nerve signs are less frequent.

The slow growth of these tumours allows the adaptation of the brain in a surprising degree when one sees at necropsy the real size of these lesions. At operation also the wide extension of these tumours is evident.

The clinical separation of definite syndromes is very difficult, and in our opinion it is better to unify all the epidermoids of the base of the brain in one single group, although the extension into different areas may induce various groups of signs.

Finally, the long histories of these cases may lead to wrong neurological diagnosis, such as trigeminal neuralgia, multiple sclerosis, etc.

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