

# LIPOMAS OF THE PINEAL REGION

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## BACKGROUND

Intracranial lipomas are rare lesions, representing 0.02% of operated intracranial lesions. Lipomas of the pineal region are even rarer and are reported occasionally in the literature. This is possibly because of the fact that they may give rise to clinical symptoms rather infrequently. When they become symptomatic, some form of management must be contemplated.

## METHODS

Four cases of lipomas of the pineal region observed in our institute during a 5-year period were investigated, and clinical-diagnostic features were studied. Two of the cases were symptomatic and were submitted to direct surgical treatment.

## RESULTS

Computed tomography (CT) scanning was performed in 3 cases and magnetic resonance imaging (MRI) in all 4. MRI angiography was performed in the last case. The appearance of the lipoma was quite pathognomonic in the neuroimaging diagnostic test. The infiltrative character of the lesion was better defined by MRI. MRI angiography gave evidence of the upward displacement of the deep veins. Direct surgical approach was performed via a right occipital transtentorial approach in one case and supracerebellar route in another. Neither approach appeared to be superior to the other one. Total removal of the lesion seemed to be impossible because of the infiltrative character of the lesion. Postoperative results were satisfactory even with incomplete removal of the lesion.

## CONCLUSION

Lipomas of the pineal region are rare. Modern neuroimaging permits a straightforward diagnosis of the nature of intracranial lipomas in general and of these lipomas in particular.

Some form of management must be contemplated when these lesions become symptomatic. Direct surgical approach, either via an occipital transtentorial or an infratentorial supracerebellar approach, is feasible. Tumor removal must be dealt with cautiously because total removal is impossible without unacceptable postoperative deficits, and generous partial removal warrants long-term symptomatic improvement. © 2004 Elsevier Inc. All rights reserved.

## KEY WORDS

Intracranial lipoma, pineal region, CT, MRI, surgical management.

Intracranial lipomas (ICL) are rare and incidentally discovered benign lesions, occurring equally in both sexes. Usually they are clinically silent, and this explains the difficulty of a proper statistical assessment. According to the largest surgical reviews, ICL represent approximately 0.02% of intracranial operated lesions [16], but considering only the neuroradiological series, their incidence becomes significantly higher – 0.08% [4,9].

The pineal-quadrigeminal plate region is not a very uncommon site of intracranial lipomas (13–26%), corpus callosum being the most frequent location [1,3,6,9,11,13–16]. However, reports on lipomas located close and/or on the pineal region are scarce, resulting from either the rarity itself of these lesions and/or to the extreme rarity of operated lipomas in the above mentioned location [2,11].

We recently observed 4 cases of lipomas located in the pineal region, 2 of which were submitted to surgery with the aim of alleviating symptoms that were considered related to the lipoma, and resulting from the pressure extended by the lesion on the surrounding structures.

## CLINICAL MATERIAL AND METHODS

In the years covered by the present study (1994–1998), approximately 20,000 neuroimaging cranial investigations were performed in the Institute of Neurosurgery "N.N.Burdenko" (18,000 cases) and in the "Clinica Nuova Latina" (2,000 cases).

The Clinica Nuova Latina is a 72-bed private hospital, credited by the Italian National Health System, with strong interest for diagnosis and management of diseases of the nervous system.

Out of this case material, 17 cases were diag-

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**1** Clinical Features

	SEX	AGE	SYMPTOMS	DURATION OF LATENCY <sup>a</sup>
Case 1	M	21	occasional headache	—
Case 2	F	10	occasional strabismus and ptosis <sup>b</sup>	—
Case 3	F	42	headache occasional vomiting, mild ataxia	3 years
Case 4	F	58	ataxia, "centro encephalic" epilepsy	1 year

<sup>a</sup>From first symptom to operation.<sup>b</sup>Confirmed diagnosis of myasthenia gravis.

nosed as intracranial lipomas, 14 in Moscow and 3 in Rome, and 4 of these cases were located in the pineal-quadrigenal region. These cases represent the basis of the present report.

**CLINICAL CHARACTERISTICS**

They were 1 male and 3 females whose age ranged from 10 to 58 and averaged 33.9 years. Table 1 summarizes the clinical characteristics of the present case material.

In 2 cases, the lesion was detected occasionally during investigations for previous head trauma. One case complained of headache and occasional dizziness of a few years duration, and these symptoms could not be related unequivocally to the radiologically diagnosed lesion. In Cases 3 and 4 the lipoma was considered symptomatic, and the patients submitted to surgery.

**CASE 3**

A 42-year-old woman was admitted in the Burdenko Institute for evaluation of her symptoms of mild,

progressively increased intracranial pressure (ICP). Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated a 2 × 2.5 cm lipoma located in the quadrigenal region, partially occluding the Sylvian aqueduct and causing remarkable hydrocephalus (Figure 1A,B). On neurologic examination a mild ataxia was also noticed.

Because of the progression of the clinical history, as well as presence of hydrocephalus, surgery was indicated. Via a right occipital transtentorial approach the lipoma was exposed in the quadrigenal plate. Debulking was performed with ultrasonic surgical aspirator and bipolar coagulation. While removing the tumor on the right side close to its capsule, the IV nerve was inadvertently cut. Therefore, removal of the contralateral portion of the lesion was intentionally very cautious and incomplete (Figure 2). After surgery, hydrocephalus persisted and had to be treated with endoscopic third ventriculostomy. Ataxia and headache definitely improved subsequently. To our surprise, the expected postoperative right-fourth nerve palsy cleared completely within 2 months.



**1** (A,B) MRI, sagittal views. T1 (A), and T2 (B) images show a 2 × 2.5 lipoma in the quadrigenal cistern. Note the reduction in the diameter of the Sylvian aqueduct and consequent significant ventricular dilatation.



**2** Postoperative MRI, T1 imaging, sagittal view. Partial (average 60%) removal of the lipoma is well demonstrated. The diameter of the Sylvian aqueduct appears to be a little wider if compared with preoperative imaging; however, hydrocephalus persists.

**CASE 4**

This 58-year-old woman underwent a cranial MRI for mild cerebellar ataxia and occasional epileptic fits occurring in the last year. Electroencephalogram (EEG) showed a "centro-encephalic" type of epilepsy, with slow waves and occasional spikes on the temporo-occipital areas bilaterally, occurring with hyperventilation. MRI showed a 3.5 × 3.5 cm lipoma located in the quadrigeminal region, slightly

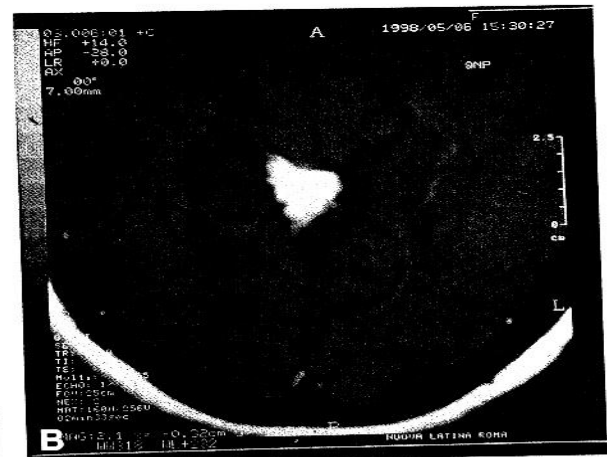
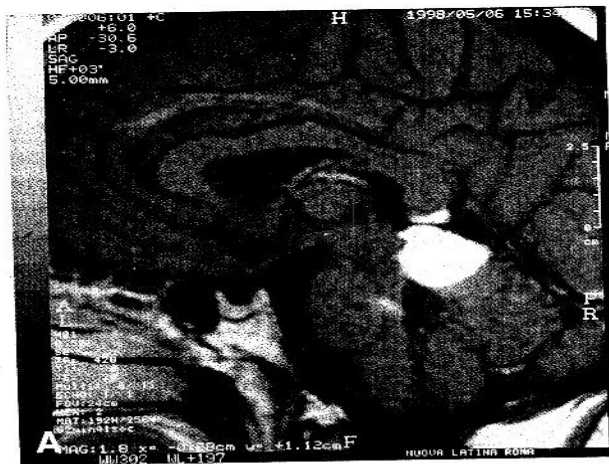
compressing the inferior aqueduct but not causing hydrocephalus (Figure 3). Decision was made for surgery since the patient, a nurse, was definitely incapacitated in her working activity by the cerebellar symptoms, while the possibility of epileptic fits being related to the lesion was considered unlikely, although it was not excluded. At surgery, a generous partial-subtotal removal (80.85%) was performed via an infratentorial-supracerebellar approach (Figure 4A,B). There was a temporary (1 month) downward gaze paresis on the right. Other than that, the postoperative course was uneventful. Cerebellar ataxia was only mildly improved; however the patient could resume part-time work. Epileptic fits disappeared, and the last control EEG appeared to be unremarkable.

**DIAGNOSTIC NEUROIMAGING**

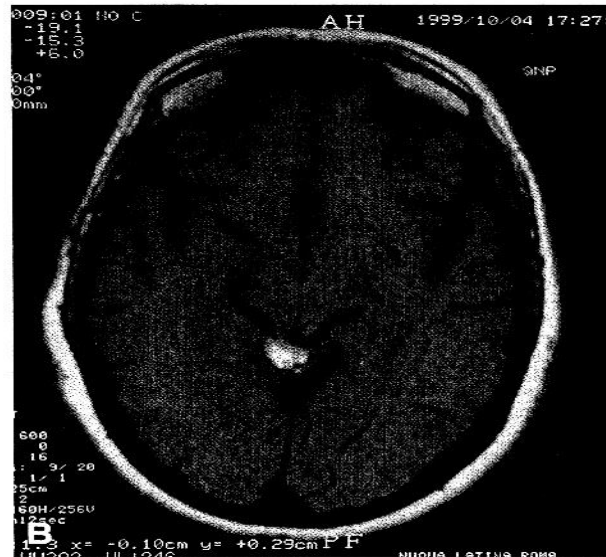
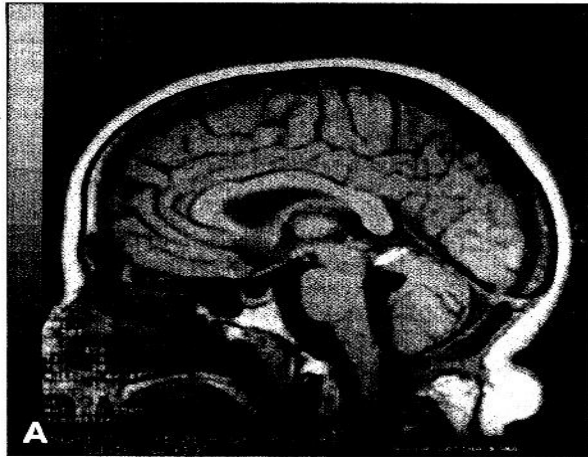
Plain X-rays were unremarkable in all cases. CT was performed in Cases 1 to 3, and considered unnecessary in Case 4. As a rule, CT scans showed a highly hypodense (H.U. I - 70 on average) lesion of variable shape, apparently well defined from the surrounding nervous structures (Figure 5).

MRI was performed in both T1 and T2 relaxation time in all cases. One case had also post-gadolinium MRI scanning. In all cases, the T1 lesion appeared highly hyperintense in T1 while intensity decreased in the T2-weighted examination, and did not enhance with paramagnetic contrast. The infiltrative character of the lesion was better defined by MRI scan (Figures 6 and 7).

MRI angiography, performed in Case 5, showed



**3** (A,B) MRI, T1 imaging, sagittal views (A) and axial (B). A large (3.5 × 3 cm) lipoma is evident. The slight compression of the inferior aqueduct does not cause hydrocephalus.



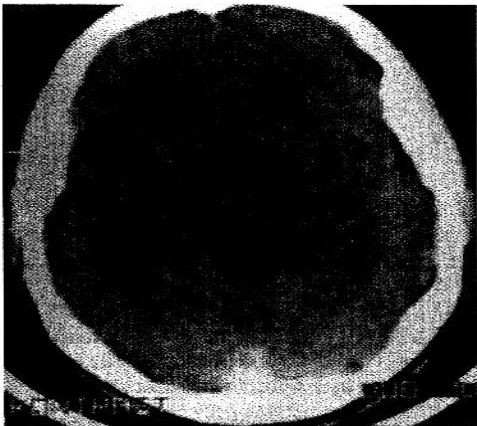
**4** (A,B) Postoperative MRI, T1 imaging. Sagittal (A) and axial (B) views give evidence of generous partial-subtotal (average 85%) removal of the mass.

evidence of upward displacement of the deep veins (Figure 8).

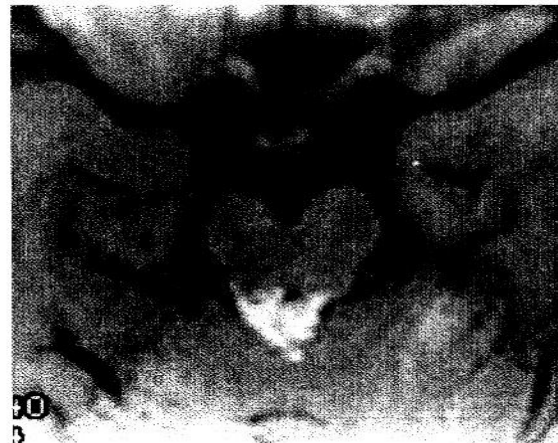
#### MANAGEMENT POLICY

The asymptomatic case was not recommended for surgery, nor was the case whose symptoms could not be certainly attributed to the lesion. In Cases 3

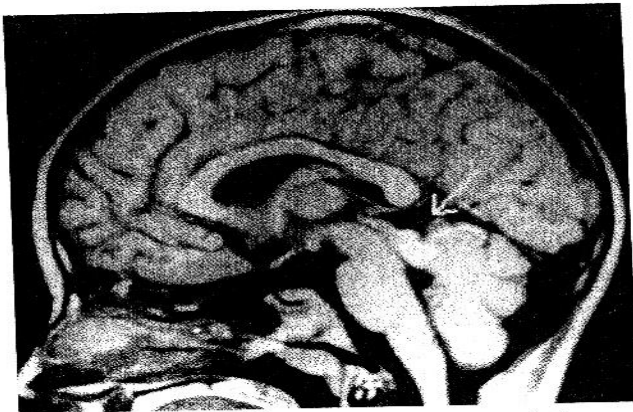
and 4 a decision for surgical treatment was made. Reasons included considerable size of the offending lesion, symptoms related to local pressure (upward gaze paresis and cerebellar ataxia), hope to relieve impending hydrocephalus in Case 3, and to positively influence the late-occurring "centroencephalic" epilepsy in Case 4.



**5** Case 1, CT scanning shows a highly hypodense lesion in the quadrigeminal cistern, eccentrically located on the left.



**6** Case 3, T1 MRI scanning, axial view. The infiltrative character of the lesion is well demonstrated.



**7** Case 2, T1 MRI scanning, sagittal view. A lipoma infiltrating the superior calliculum and the upper vermis is well shown.

### SURGICAL APPROACH

Case 3 was operated in Moscow using an occipital transtentorial approach. The angle of vision obtained by sectioning the tentorium was quite adequate for visualizing the margins of the lesion from above, although the surgeon had to work sometimes very close to the deep veins that were dislocated upwards. However, the adequate vision did not prevent inadvertently injuring the right trochlear nerve, which was embedded in the tumor capsule and was visualized only when sectioned. Therefore, the removal of the contralateral portion



**8** Case 4, MRI angiography. This examination shows upward displacement of the deep vermis.

of the lesion was intentionally not radical, for fear of also injuring the left trochlear nerve. At the end of surgery, removal was thought to be generous-partial ( $\pm 60\%$ ), and this was confirmed by postoperative neuroimaging (Figure 2). Partial removal did not allow us to relieve pressure to the Sylvian aqueduct. Therefore, an endoscopic III ventriculostomy was planned and successfully performed 2 months later.

Case 4 was operated in Rome using an infratentorial suboccipital approach. The lesion was easily separated from the deep veins and the superior cerebellar arteries. However, as expected, adequate visualization of the ponto-mesencephalic junction was limited by the angle of approach. Therefore, no attempt was made to remove the lesion too aggressively anteriorly for fear of injuring the trochlear nerves. Nevertheless, during the removal of the last piece of tumor, a small vessel coming from the right superior cerebral artery (SCA) was injured and eventually coagulated; removal was thought to be generous-subtotal (80-85%), and this also was confirmed by postoperative MRI.

### RESULTS

Case 2 (not operated on) is stable 5 years following discovery of her lesion, and she is being followed with observation.

As stated above, either generous subtotal or partial removal in surgically treated cases led to improvement of the preoperative clinical symptoms. Disappearance of epileptic fits in Case 4 matched well with normalization of control EEG. Both cases suffered from postoperative unilateral trochlear nerve palsy (in 1 case expected). As stated above, this cleared within 1 month postoperatively in both cases.

### DISCUSSION

#### GENERALITIES OF INTRACRANIAL LIPOMA

Intracranial lipomas are considered to be congenital lesions [3,7], most likely related to maldevelopment of the "meninx primitiva," which is a mesenchymal derivation of the neural crest [12,13].

These lesions are rare. Their actual incidence approximates 0.02% of operated intracranial lesions in very experienced hands (16), and appear to be higher (0.08%) purely in either autoptic [3,15] or neuroradiological series [4,9]. This apparent discrepancy is well explained by the clinical indolence of the largest part of the intracranial lipomas, very often incidentally discovered. Nevertheless, they

can exert pressure on local structures, thus becoming symptomatic in some instances.

The mechanism with which they cause symptoms is not clear. While it is widely accepted that they may grow until individual statural growth terminates, it is also clear that their growth does not necessarily result in the lesions becoming symptomatic [9]. There is no proof in the literature that intracranial lipomas grow and later become symptomatic because of its growth [2]. Therefore, the production of clinical symptoms because of local pressure has been related to hypothetical slow growth by some [2,6] while others [9] have suggested that regressive changes occurring within the lipomatous tissue might secondarily exert traction, thus affecting the surrounding nervous structures.

Whatever is the cause, there is general agreement that when the lesion appears to be symptomatic, some form of management must be contemplated [9].

### DIAGNOSTICS OF INTRACRANIAL LIPOMAS

CT scanning and MRI examination represent the main diagnostic tools at the present time for intracranial lipomas [5,9,10]. Their appearance on these investigations is quite pathognomonic and obviates the need for diagnostic biopsy in virtually every case.

At CT scanning, intracranial lipomas appear as homogeneous lesions of extremely low attenuation value ( $-70$  H.U. on average), sharply demarcated from the surrounding neural structures and not enhancing with contrast [2,4,9,10]. Calcifications may on occasions be present, usually at the periphery of the lesion [9]. MRI examination of intracranial lipomas, as a rule, shows a homogeneous lesion hyperintense on T1 and hypointense on T2, with short relaxation times [5,13,14].

CT and MRI scanning features are generally considered pathognomonic of intracranial lipomas [2], and as a rule they help establish a preoperative diagnosis.

### LIPOMAS OF QUADRIGEMINAL PLATE

Lipomas of quadrigeminal plate represent more than 10% of intracranial lipomas [2,9,11,13]. However they are usually asymptomatic, thus, are found occasionally during neuroimaging investigation for unrelated clinical problems.

Reviewing 203 cases of intracranial lipomas from the international series, Maiuri et al [11] found that only approximately 20% of lesions located in the quadrigeminal—pineal region give rise to clinical symptoms, possibly related to local pressure. Al-

though this would appear to be an infrequent evidence, there are convincing reports in the literature that lipomas of the quadrigeminal-pineal region can exert pressure on the surrounding structures, thus causing clinical symptoms [2,9,11].

Furthermore, lipomas located in this critical location can sometimes be symptomatic [1,2,6,9], even if they did not attain a significant size [8].

### MANAGEMENT POLICY

Patients harboring intracranial lipomas that are either asymptomatic, or show symptoms not clearly and definitely attributable to the lesion should be managed with observation. This also applies to lipomas of the quadrigeminal plate, and this was our experience in Cases 1 and 2. When the lesion is symptomatic, some form of treatment must be contemplated. Baesa et al [2] recently reviewed the literature on this topic and found only 8 cases of lesion of the quadrigeminal plate which had been submitted to surgery, 4 of these cases were treated only with a shunting procedure because of hydrocephalus being a prominent clinical feature. In our Case 3 we decided to submit the patient to direct surgery, although hydrocephalus was the main clinical feature, because a shunt would not have resolved the associated, though minor, clinical problem such as ataxia.

As in the Case 1 of Baesa et al [2], extensive partial removal resulted in relieving the cerebellar preoperative dysfunction. However, hydrocephalus did not improve significantly, and the patient later had to undergo a form of cerebrospinal fluid (CSF) diversion. Intraoperative injury of the right IV nerve prevented us from attempting a (sub)total removal of the lesion, which we planned preoperatively for decompressing the Sylvian aqueduct and relieving the preoperative hydrocephalus. Interestingly, verified unilateral trochlear nerve injury was compensated without further treatment within a few weeks.

Our Case 4 was operated on, again with the hope of relieving cerebellar symptoms. Interestingly, she suffered from infrequent attacks of "centro-encephalic" epilepsy, which cleared after subtotal removal. Both cases were operated on using microsurgical techniques, with extensive use of bipolar coagulation and careful removal of lipomatous tissue using microinstruments. However, as stated above, this did not prevent injuring the trochlear nerves in Case 3, and a branch of SCA in Case 4, which fortunately did not result in any clinical postoperative worsening. This means that no attempt should be made to work aggressively on the peripheral part of the lipoma, because this usually can be intimately adherent to critical structures such as

arteries, nerves and the mesencephalon. Lesion removal should be very accurate, and we would not recommend using aggressive surgical tools such as ultrasonic aspirator in the proximity of the infiltrating portion of the lipoma.

The surgical approach was different in the 2 cases. Although the angle of vision obtained with the supratentorial approach is theoretically better, this did not obviate the technical difficulties related to the deep and narrow surgical field, and the necessity of working close to the deep veins.

Subtentorial approach better exposes the posterior portion of the tumor; however, it requires a significant downward retraction of the culmen to properly visualize the tumor, and does not offer any chance to see the anterior and inferior portion of the lesion, on which the surgeon is forced to work with some blind maneuvers. Extensive partial-subtotal removal of these lesions being the surgical goal, either of these approaches appears to be appropriate, as both have equivalent advantages and shortcomings. A combined supra-and subtentorial approach, as described by Ziyal et al [17] for falco-tentorial meningiomas, is not to be recommended, in our opinion, because it is very demanding technically and lipomas are as a rule not totally resectable because of their infiltrative character.

## CONCLUSIONS

Intracranial lipomas are rare, and rarer are lipomas of the pineal region. They can produce clinical symptoms usually by exerting local pressure on dorsal mesencephalic structures and on the Sylvian aqueduct. Indication for direct surgery should be posed with caution, and the advantage of relieving local pressure should be balanced against the risk of approaching the pineal region, in line with the personal experience of the operating surgeon. In cases where hydrocephalus is a major feature, a shunting procedure can be a sound alternative. As for a direct surgery, either approach (supra- or subtentorial) seems to appropriate in facilitating a near-total removal of the lesion, while a generous partial-subtotal removal would also appear to produce satisfactory postoperative results.

The ability of dorsal mesencephalic structures to compensate unilateral IV nerve injury—as suggested by experience with our first operated case—deserves further attention, but should not be a reason for suggesting more aggressive surgical conduct in removing infiltrative brain tumors involving the collicular region.

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## COMMENTARY

Because little attention is given to cerebral lipomas, especially for lipomas of the pineal region, this presentation of 4 cases provides an important glance to this rare condition, which might be encountered eventually by most neurosurgeons despite the rarity of this pathology. In a personal series of 115 cases of pineal region tumors only 2 cases (1.7 %) had been observed, the percentage slightly higher

as discussed here from the literature. Lipomas have a low Hounsfield number ( $-70$ ), and in combination with their slightly lobulated mass with no capsule-like border, allow a presumptive histological diagnosis in CT-imaging alone. Differential diagnosis with other cystic lesions of other histological nature should be possible by the enhancement of their capsule of the latter by contrast material in CT or MRI. The authors stress the impossibility of a total resection and that only a partial resection should be advocated, if surgical approach is indicated at all. In our own 2 cases [1] with mild clinical signs and symptoms besides aqueductal occlusion, partial resection brought relief of these clinical fea-

tures, but insufficient evacuation of the hydrocephalic pathology and shunting procedure followed. Therefore, we agree with the authors that in cases of lipomas of the pineal region with only aqueductal stenosis or occlusion and with clinical signs of hydrocephalus, a shunting procedure should be the only treatment of choice.

**Prof. Gerhard Pendl**  
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**A** religious man is a person who holds God and man in one thought at one time, at all times, who suffers harm done to others, whose greatest passion is compassion, whose greatest strength is love and defiance of despair.

—ABRAHAM JOSHUA HESCHEL,  
JEWISH THEOLOGICAL SEMINARY OF AMERICA, NYC