

# Once in a lifetime: a case of a pineal tumour

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**SUMMARY.** This paper describes the case of a 14-year-old boy who presented to his general practitioner with a history of headaches and visual problems. Investigations subsequently revealed that he had a pineal tumour. The management of the patient is described and this rare condition discussed.

## Presentation

A 14-YEAR-OLD boy, accompanied by his father, presented at their general practitioner's surgery on 7 May 1985. His initial complaint was of persistent headaches, which had become worse over the previous six weeks and were waking him in the mornings. On direct questioning it transpired that the headaches had begun around Christmas 1984. At the time he remembered developing a headache after heading a football. He also experienced difficulty playing snooker as he could not raise his eyes. The headaches became worse and more frequent, and were associated with nausea.

The family went on holiday to Majorca in February and all of them suffered a viral-like illness, but the patient's headache and vomiting lasted longer than the rest of the family. This was followed by increasing lethargy, dizziness and difficulty focussing. Following a temporary improvement in his symptoms, he was brought to the surgery when his headaches had become far worse and his father had also noticed a change in personality.

On examination, the patient looked subdued and unwell. His pulse rate was 55 per minute and blood pressure was 140/85 mmHg. The pupils were largish, reacted to accommodation but slowly to light. The boy was unable to look upwards and there was gross bilateral papilloedema.

## Management

A provisional diagnosis of raised intracranial pressure was made, and the patient was immediately admitted to the local hospital paediatric ward. A computerized tomography scan performed later that day (Figure 1) showed a symmetrical midline mass, 3cm in diameter, around the pineal calcification, which was noted to be unusually extensive for a person of this age. The scan also showed gross symmetrical hydrocephalus of the lateral and third ventricles. The appearances were considered typical of a pineal region neoplasm, probably arising from the pineal gland.

The patient was then transferred to the regional neurosurgical unit. The following day he underwent a right ventriculo-peritoneal shunt. No malignant cells were seen in the cerebrospinal fluid. He was eventually discharged home, well, on 17 May 1985. He then had a course of central nervous system radiotherapy to the spine, the whole brain and additionally to the pineal region using a cobalt unit. Following this, the patient had persistent vomiting and was readmitted from home back to the local paediatric ward for reassessment. A repeat computerized tomography scan (Figure 2) on the 7 August showed the tumour and ventricles to be much smaller. A diagnosis of post-radiation sickness was made which eventually responded to treatment with domperidone. The patient was allowed home two days later and went back to school in September.

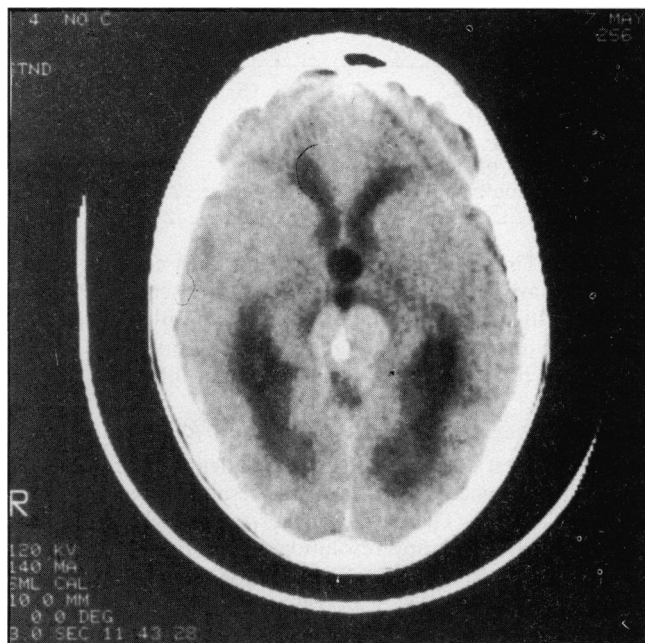


Figure 1. Computerized tomography scan taken on day of presentation, showing the calcified pineal gland and surrounding pale area representing tumour.

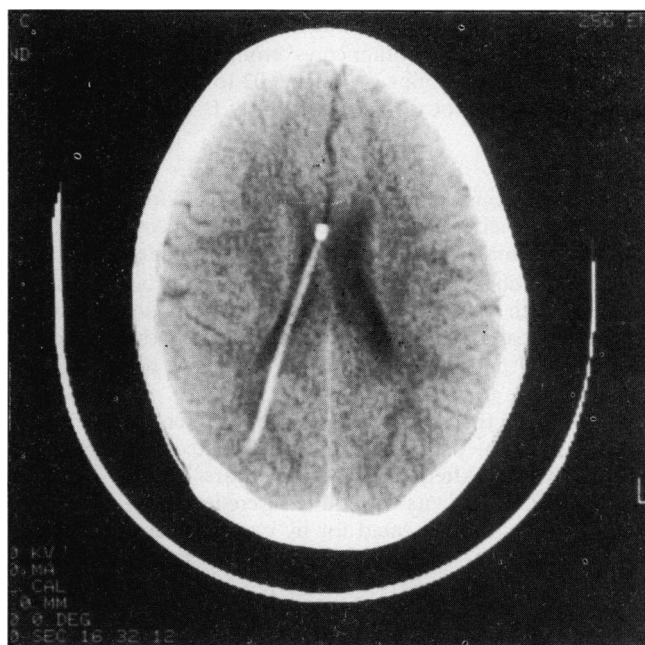


Figure 2. Computerized tomography scan taken three months later after insertion of shunt and radiotherapy, showing normal sized ventricles and shunt in position.

At this early stage, the prognosis of the case described must still be guarded. Nevertheless, at the time of writing, the boy concerned has been back at school full time for two terms.

## Discussion

Primary brain tumours account for a mere 2% of all deaths from malignant disease.<sup>1</sup> A general practitioner with a list of 2500

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patients will see, on average, one new case of brain tumour every 10 years.<sup>2</sup> Of these, tumours of the pineal gland make up a very small proportion.<sup>1</sup> Nevertheless, the pineal gland has always held a strange fascination for doctors, biologists and philosophers alike. Descartes called it 'the seat of the soul'.

The pineal gland is a small body attached by a stalk to the roof of the third ventricle. It is maximally developed by about seven years of age then slowly regresses. In 60% of adults it is calcified sufficiently to be seen on skull radiographs, and its position can be helpful in diagnosing lateral displacement of the cerebral hemisphere due to space-occupying lesions. The function of the normal pineal gland remains uncertain, but it appears to have a secretory role, possibly of melatonin. In lower vertebrates such as the frog, the pineal contains photoreceptive cells.

Tumours of the pineal gland are very rare, but when they occur are most commonly found in children and young adults. They are also more common in males than females. Probably the first documented case of a pineal tumour was by Charles Drelincourt, published in 1717 in Geneva.<sup>3</sup> During the nineteenth, and the first half of the twentieth century, owing to the variety of histological types, many names were applied to tumours of this region.<sup>4</sup> Today, most tumours of the pineal region are classified as either germinal (non-parenchymal) or pineal parenchymal in origin. The former are the more common, including dysgerminomas and teratomas. In less than 10% of cases these have been linked with precocious puberty in young boys caused by tumour-secreted human chorionic gonadotrophin or gonadotrophin.<sup>5,6</sup> The pineal parenchymal tumours include the pineocytoma, which is usually benign and the malignant pineoblastoma, which resembles neuroblastomas, medulloblastomas and even retinoblastomas, where the cells show similarities with the specialized site of origin.<sup>7</sup> Parenchymal tumours may be associated with hypogonadism, possibly caused by a reduction in the secretion of pineal anti-gonadotrophic factor.<sup>8</sup>

The treatment of pineal tumours is preferably by radiotherapy as surgery has a high morbidity and mortality, and germinomas are highly radiosensitive.<sup>9,10</sup> Radiotherapy can be curative, but in cases with raised intra-cranial pressure, a shunt may be required as well. The advent of computerized tomography has made this method of management (cerebrospinal fluid shunting and radiotherapy) justifiable as a trial therapy. Even biopsy of these tumours is hazardous because of the risk of haemorrhage. Prior to computerized tomography, ventriculography was necessary to outline (incompletely) the tumour and only hint at its size and any change in size produced by the radiotherapy.

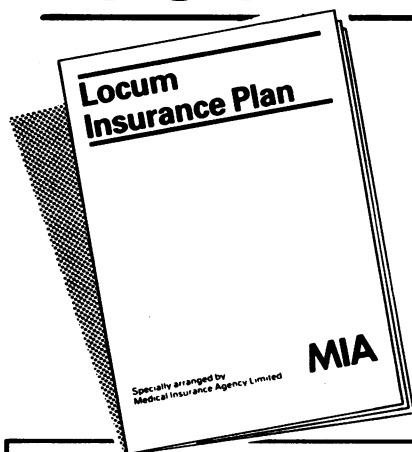
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