

# Delayed intra-tumoural haemorrhage in pineal germinoma: Case report and review\*

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## ABSTRACT

**Intraparenchymal haemorrhage in a pineal germinoma is a very rare, though clinically significant event. We report the first case of a significantly delayed intraparenchymal haemorrhage in a pineal germinoma, 14 days after endoscopic third ventriculostomy (ETV), causing precipitous patient deterioration. We discuss potential contributing pathophysiological factors, and seek to illustrate that knowledge of its occurrence, and associated morbidity, is clinically significant in managing pineal germinoma patients with acute deterioration post obstructive hydrocephalus CSF diversion.**

**Keywords:** Pineal Germinoma; Haemorrhage; Endoscopic Third Ventriculostomy (ETV); Tumour

## 1. INTRODUCTION

Pineal germinoma is a rare primary brain tumour (incidence 0.4% - 3.4% [1]) that classically presents with obstructive hydrocephalus or local mass effect [2]. Initial management involves treating the hydrocephalus with either endoscopic third ventriculostomy (ETV) or a shunt procedure, and gaining a tissue diagnosis to direct definitive treatment. Haemorrhage within pineal tumours is only rarely reported as a very early, but clinically significant, complication of treatment. We describe a very late and significant intraparenchymal haemorrhage with ventricular extension from a pineal germinoma, delayed at 14 days after ETV and biopsy.

## 2. CASE REPORT

A 15-year-old male presented with a four week history of progressive morning headache, nausea, vomiting, and

diplopia.

Examination revealed paralysis of upward gaze, convergence-retraction nystagmus, and pseudo-Argyll-Robertson pupils consistent with Parinaud's dorsal midbrain syndrome, partial bilateral abducens nerve palsy, and papilloedema.

MRI brain demonstrated a large homogenous enhancing pineal tumour with prominent vascularity and associated obstructive triventricular hydrocephalus.

An ETV with tumour biopsy was performed; minor haemorrhage was noted intra operatively at the biopsy site which resolved quickly with irrigation. An extraventricular drain (EVD) was inserted as a precaution, and removed 24 hours later. The patient recovered well, with a significant improvement in the majority of his symptoms however the Parinaud's syndrome was slower to resolve. Histology, and elevated CSF beta-human chorionic gonadotropin and alpha fetoprotein levels, confirmed pineal germinoma.

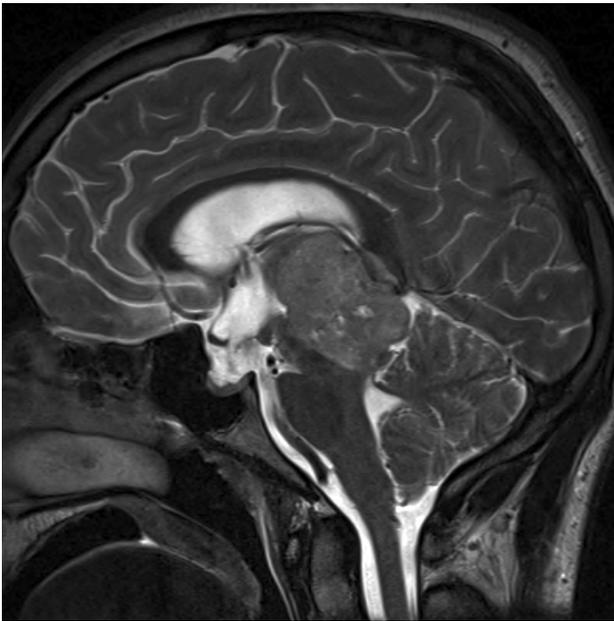
Post-operative MR (**Figure 1**) confirmed patency of the third ventricle stoma, significant reduction in ventriculomegaly, with no intraventricular or intratumoural haemorrhage.

A CSF leak developed from the right frontal EVD site which resolved with simple re-suturing and 50 mL of CSF removed via lumbar puncture. Given the patency of the stoma it was considered possible that an element of communicating hydrocephalus may have developed. The patient remained clinically very well, until a sudden drop in conscious level to GCS 8 on day 14, accompanied by urinary incontinence and emesis. The patient was intubated and emergent CT imaging (**Figure 2**) was performed, which revealed a large diffuse parenchymal haemorrhage within the tumour, intra-ventricular haematoma and acute hydrocephalus.

A right frontal EVD was reinserted immediately. Post-EVD CT imaging revealed a dilatation of the left ventricle secondary to castes of blood situated both at the fo-

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**Figure 1.** T2 sagittal MRI brain: Two days post ETV. Third ventricle stoma patent.



**Figure 2.** CT contrast brain: Diffuse enlargement of pineal tumour secondary to large intraparenchymal haemorrhage with intraventricular extension and acute hydrocephalus.

ramina of Monroe and overlying the third ventricle stoma. This prompted placement of a left frontal EVD.

The EVDs were gradually weaned and chemotherapy commenced to reduce the tumour bulk. The patient was

discharged home a fortnight later with considerable reduction of symptoms, and serial imaging confirming progressive reduction in tumour size.

### 3. DISCUSSION

Intra-tumoural haemorrhage in pineal germinomas is a rarely reported complication following treatment of obstructive hydrocephalus by either ETV or shunt procedure. Large clinical studies assessing ETV complications do not describe this specific type of post ETV haemorrhage [3-7]. It has however been reported, only in the acute setting, in a small number of individual case reports following CSF diversion for pineal tumours without biopsy. The presentations varied from a small asymptomatic bleed [8], acute deterioration at two hours [9] and on day two [10], and a fatal bleed at 5 hours [11]. Clinically significant haemorrhage post-CSF diversion for obstructive hydrocephalus caused by other posterior fossa tumours, again without biopsy, has also been described [12-19].

It has been hypothesised that altered CSF dynamics post CSF diversion is the major precipitating event causing tumour haemorrhage [9]. Proposed pathophysiological mechanisms implicate the acute reduction of CSF pressure in one ventricular compartment resulting in an increased transmural pressure gradient across the tumour, with progressive distortion of fragile tumour vessels. Resultant tumour movement may further compress venous drainage leading to venous hypertension, increased tumour oedema and spontaneous parenchymal haemorrhage [12,20]. All clinically relevant tumour haemorrhages reported have occurred by Day 2, which is consistent with the proposed mechanism of haemorrhage secondary to acute changes in intracranial pressure (ICP). The only other reported case of pineal haemorrhage was a clinically insignificant bleed at Day 10, only incidentally noted on preoperative imaging [8].

The aetiology for such a delayed and clinically significant tumour haemorrhage in our patient at Day 14 is unclear. Although minor haemorrhage followed the endoscopic biopsy (a well-recognised phenomenon), post-operative imaging confirmed that this was not significant. We do not feel that the diffuse nature of the haemorrhage in our patient related to either the biopsy or to the acute changes in ICP responsible for the reported clinically significant haemorrhages.

The possible development of an element of communicating hydrocephalus (albeit mild) may have resulted from the initial biopsy haemorrhage. Persistently elevated ICP may have contributed by exacerbating venous congestion, leading to tumoural microhaemorrhage, oedema, venous infarction and final large haemorrhage [21].

Pineal germinomas occasionally present with a spon-

taneous bleed causing subarachnoid haemorrhage [21-24]. Particular tumour features associated with spontaneous haemorrhage [24] may have predisposed this prominently vascular, and untreated, tumour to haemorrhage independent of ETV effect.

Whether the cause of the haemorrhage is related to altered CSF dynamics, transmural mechanical stress, vascular congestion, elevated ICP, or simply stochastic, remains equivocal. It is, however, a previously undescribed phenomenon, that is a clinically important differential for the patient with delayed-acute deterioration.

#### 4. CONCLUSION

Pineal germinoma haemorrhage post ETV is a rare and clinically relevant complication, previously described as occurring only in the acute setting. We report the first case of a haemorrhage occurring in a very delayed fashion (Day 14) with resultant intraventricular haemorrhage, and hydrocephalus (despite ETV) that caused significant acute patient deterioration. This significant complication has associated morbidity, and knowledge of its occurrence deserves consideration and action from the surgeon managing patients after CSF diversion for obstructive hydrocephalus in the setting of a pineal germinoma.

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