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A Case Report of Pineal Germinoma Apoplexy

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1. Abstract

1.1. Background

Pineal apoplexy is a condition related to the pineal gland of the brain, characterized by bleeding within this region. The bleeding can be caused by various factors, including pineal cysts, tumors, or abnormal blood vessels near the pineal area. Intracranial germinomas (GN), rare types of tumors that typically start in children, can also be a factor.

1.2. Case Presentation

A 16-year-old boy who had previously been diagnosed with a pineal gland tumor (Germinoma) and was undergoing chemotherapy presented to our hospital with complaints of headache and diplopia. The patient has past history of intra ventricular CSF diversion to relive high intracranial tension. An urgent CT scan revealed a soft tissue mass in the pineal region, accompanied by supraventricular hydrocephalus, intraventricular hemorrhage, and CSF seeding. The patient's Glasgow Coma Scale (GCS) was 10, and he was admitted to the ICU for observation due to deteriorating vital signs.

1.3. Methods

Urgent brain CT was done with multi–detector row scanners. Multi–detector row CT images were acquired in four simultaneous 2.5-mm-thick sections, which were combined in projection space to create two contiguous 5-mm-thick sections. Axial, sagittal and coronal reconstructions non contrast CT was done.

1.4. Results

CT scan showed a soft tissue density mass in the pineal region with hyperdense areas near the septum pellucidum and the ipsilateral right lateral ventricles. Supratentorial hydrocephalus with ependymal spread in both lateral ventricles was also detected.

1.5. Objectives

Imaging techniques like CT and MRI play a crucial role in identifying the specific cause of the pineal apoplexy.

2. Introduction

The pineal gland is a sophisticated part of the body that can be impacted by a wide range of benign and cancerous growths [1]. Primary central nervous system germ cell tumors (CNS-GCT) are an uncommon kind of cancer that originates from germ cells situated along the midline during fetal development, primarily affecting children. It is estimated that about 90% of these tumors are found in individuals under the age of 20 [2].

Tumors in the pineal area include glandular tumors (GCTs), tumors of the pineal parenchyma, brain tumors, meningiomas, metastatic tumors from other areas, and benign growths such as pineal cysts, lipomas, and certain types of skin tumors [3].Pineoblastomas are the most prevalent tumors in the pineal area, followed by GCTs. GCTs are more prevalent among males. The World Health Organization (WHO) categorizes them into two types: germinomas and non-germinomas [4].Germinomas are primarily located in the central part of the brain, the pineal gland, and/or the areas above the pituitary gland. The symptoms of germinomas

depend on the tumor's size and location, leading to problems with hormone production, increased intracranial pressure, and changes in vision, including double vision and paranoia [5].This situation can result in acute onset of severe headaches, acute obstructive hydrocephalus, mass effect on the midbrain, and even death and this give our case report unique characters so follow up is highly recommended.

3. Case Presentation

A 16-year-old boy presented to our hospital complaining of headache and diplopia. Previous MRI had revealed a pineal mass with tiny dots of hemorrhage, and the patient was undergoing chemotherapy for 6 months. Although intracranial germinomas are highly radiosensitive and can be cured with radiotherapy alone, the current standard of care in our patient involves neoadjuvant platinum-based chemotherapy for 6 months. Due to worsening symptoms, an urgent CT scan was performed, which showed a soft tissue density mass in the pineal region with hyperdense areas near the septum pellucidum and the ipsilateral right lateral ventricles. Supratentorial hydrocephalus with ependymal spread in both lateral ventricles was also detected (Figure 1-3).

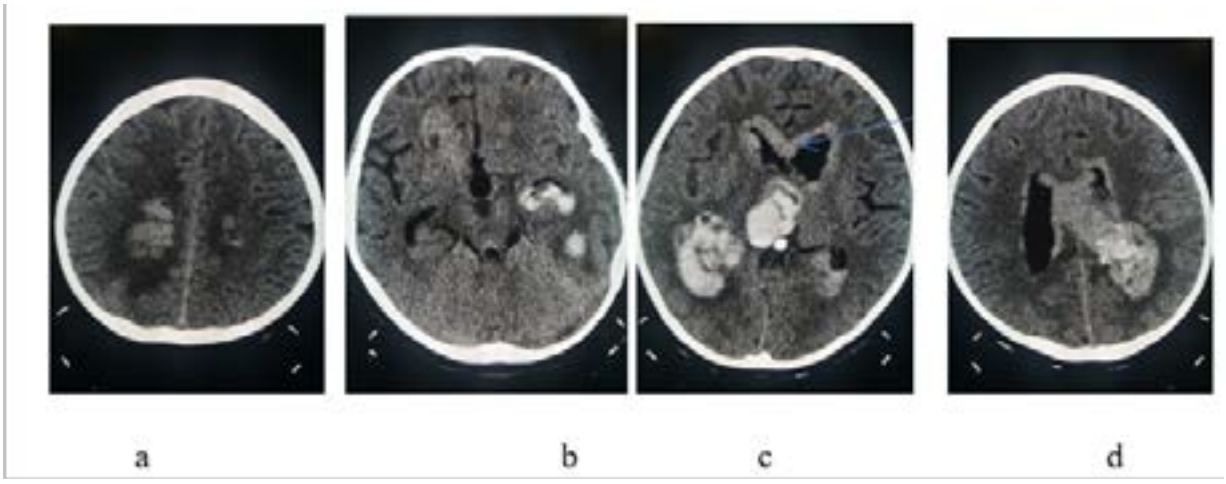


Figure (1):): Axial CT brain(a-d) images revealed hyper dense soft tissue density mass in the pineal region engulfing foci of calcification with subsequent supratentorial hydrocephalus associated with haemorrhage density inboth lateral ventricles and ependymal spread (arrow pointed)

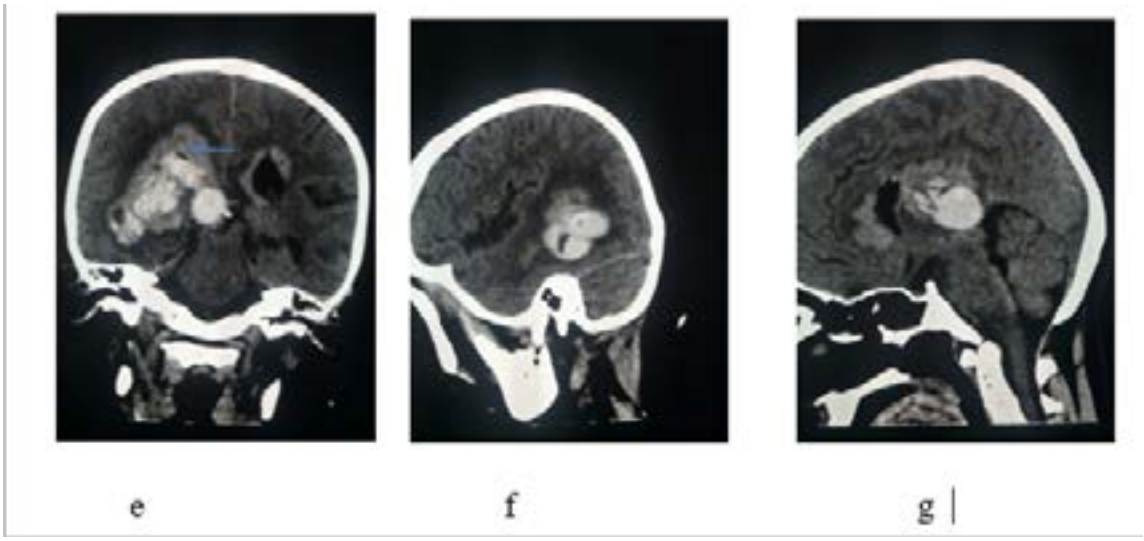
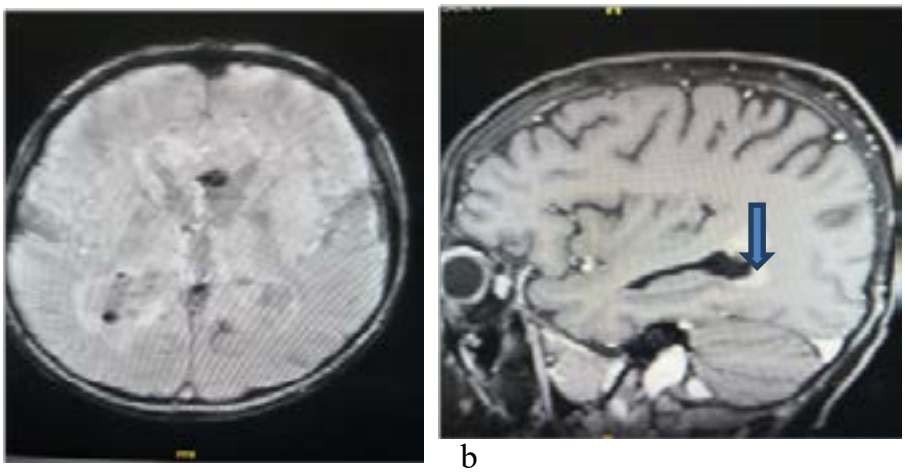


Figure (2):CT brain(e-f-g), coronal and sagittal images revealed hyperdense soft tissue density mass in the pineal region engulfing foci of calcification with subsequent supratentorialhydrocephalus associated with haemorrhage density inboth lateral ventricles and ependymal spread (arrow pointed).



a b

Figure (3): Pervious (T2*-GRE) and post contrast MRI brain(a&b) axial and sagittal sections revealed foci of hemosiderinin the pineal region and related ventricles(axial image)&Post contrast sagittal MRI revealed enhancing ependymalnodules of lateral ventricle(arrow pointed).

4. Discussion

Pineal apoplexy is an uncommon condition. Apoplexy refers to symptoms resulting from sudden intracranial bleeding in a healthy pineal area or growth [2]. Most germ cell tumors occur in patients under 20 years of age, mostly in adolescents, and are common in boys. Bifocal tumors in the pineal and suprasellar regions, even though they may appear to be so, are not considered metastatic [2].It is marked by combined signs such as headache, nausea, and vomiting. The condition is caused by blocked-off hydrocephalus and/or physical pressure on the cerebellum, midbrain pretectum, or tectum area [5]. However, it has not been documented for pineal apoplexy occurred in a normal gland. Pineal apoplexy, which involves sudden bleeding, can start in a pineal cyst, tumor, or nearby blood vessel abnormality [6].The average age of individuals diagnosed with intracranial germinoma initially is between 16 and 18 years old; 84% develop symptoms between 9 and 27 years, aligning with our situation where the patient was about 16 years old [6]. Brain tumors known as germinomas often occur above the sella turcica, in the pineal area, or in both the pineal and sella turcica. They may also occur in other areas (5%) [7].The symptoms of pineal apoplexy can be classified as paroxysmal or chronic, ranging from a few days to months or even years. The most frequent symptoms are severe frontal or occipital headache, gaze paresis and visual field defects, nausea or vomiting, syncope, and ataxia, which are due to accompanying hydrocephalus [7]. Germinomas are different from other germ cell tumors in that they do not consistently elevate serum levels of HCG or alpha fetoprotein [5]. In cases of germinomas, non-contrast CT typically reveals a sharply defined, hyperdense mass. The increased density is often attributed to intra tumoral hemorrhage. Our case demonstrates hydrocephalus with ependymal spread, a notable feature [8]. In this case, cerebrospinal fluid (CSF) analysis showed negative CSF and serum tumor markers (AFP and HCG).In patients with mild symptoms, observation is indicated, whereas surgical treatment is reserved for severe cases presenting with obstructive hydrocephalus and includes cerebrospinal fluid diversion, resection of apoplectic pineal lesions, or both [9,10]. Germinomas are very sensitive to radiation treatments, and treatment with chemotherapy and the general outlook is very good, with a 5-year survival rate around 90% [11]. Synchs tumors in the pineal and suprasellar regions are mostly not considered metastatic [12]. Follow-up MRI of the brain is mandatory after treatment, as there is a high prevalence of relapse via subependymal or CSF spread. A spinal MRI is indicated if spinal lesions were previously identified, or the patient has symptoms related to the spine [13].

5. Conclusion

Pineal apoplexy is a condition characterized by a variety of symptoms due to a sudden hemorrhage caused by bleeding in a pineal cyst, tumors within the pineal area, or abnormalities in nearby blood vessels. The severity of pineal apoplexy due to bleeding varies from no symptoms to rapid deterioration and potential death. Follow up is highly recommended as a life threatening strategy.

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