

## Adenocarcinoma of the pineal gland yolk sac tumor

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### ABSTRACT

In children, tumors of the pineal area account for 3–8% of all intracranial lesions. Pinealocytes, supporting cells of the pineal gland, or glial cells from the nearby midbrain or thalamus can cause neoplasms in this area, which can range from benign to mali-

-gnant. Up to 53% of malignant tumour's identified in the pineal area are germ cell tumour's. Malignant somatic transformation of germ cell tumour's in the pineal area is uncommon and seldom described. After multiple cycles of neoadjuvant treatment, a pineal yolk sac tumour with an enteric type of mucinous adenocarcinoma regrew relatively quickly.

**Key Words:** *Adenocarcinoma*

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### EDITORIAL

A previously healthy 15-year-old boy appeared with headaches and deteriorating mental state for two months [1]. At the time of presentation, MR imaging revealed a massive heterogeneously enhancing pineal area tumour measuring 2.3 2.0 3.2 cm<sup>3</sup> that was producing obstructive hydrocephalus. His serum Alpha-Fetoprotein (AFP) level was abnormally high (2568 ng/mL), but his Human Chorionic Gonadotrophin (HCG) level was normal. He had an endoscopic third ventriculostomy, which included a bulk biopsy and the implantation of an Ommaya reservoir. The biopsy revealed a nonseminomatous germ cell tumour, which is similar to a yolk sac tumour.

Etoposide, carboplatin, and ifosfamide were added to his chemotherapy treatment. MR imaging revealed a small decrease in the size of the tumour after 2/6 cycles of chemotherapy, and the patient's AFP level dropped to 115 ng/mL. Despite this early improvement, the patient developed severe headache, photophobia, double vision, lid lag, and an upward gaze palsy immediately after his sixth cycle, and his serum AFP level was once again considerably increased (8068 ng/mL). Furthermore, MR imaging revealed that the mass had grown in size, measuring 3.0 2.1 3.6 cm<sup>3</sup> today, with a larger mass effect on the surrounding mesencephalon. The patient underwent a right parieto-occipital craniotomy using an occipital transtentorial approach to achieve a radical subtotal resection because to radiographic progression and clinical worsening despite an initial good response to chemotherapy. The tumour was biphasic, with sections of true yolk sac tumour and areas of gastroenteric differentiation with mucinous goblet cells and irregularly shaped pleomorphic glandular components, similar to a somatic-type adenocarcinoma. The yolk sac component was positive for SALL4 and glypican-3 but negative for Monoclonal CEA (mCEA) based on

immunohistochemical staining, whereas the carcinomatous component was positive for mCEA. AFP and CDX2 were diffusely positive, SATB2 was variable, and OCT4 and CD30 were negative in both components. The overall histologic and immunohistochemical findings were compatible with a yolk sac tumour with enteric differentiation and somatic-type adenocarcinoma [2]. There were no further germ cell tumours found. While the degree of nuclear atypia in the glandular components varied, there were no benign-appearing patches to imply a monodermal teratoma. Following resection, the patient was put on an irinotecan, paclitaxel, and oxaliplatin chemotherapy regimen. Repeat MR imaging revealed additional leptomeningeal illness within a month, with distinct nodular enhancement identified at numerous levels of the cervical, thoracic, and lumbar spine [3]. At that time, it was decided to discontinue chemotherapy and begin a 6-week course of palliative craniospinal radiation. Unfortunately, the patient died from the disease about 11 months after the original biopsy and 6 months after the subtotal resection.

An intestinal kind of mucinous adenocarcinoma originating within an intracranial yolk sac tumour of the pineal region is described here. Two harmful point mutations were discovered after extensive molecular profiling. There were no glandular parts that looked like benign or mature tissue in this example, hence the presence of a teratomatous component is doubtful. Somatic-type transformation is extremely unusual in this locale, according to our understanding, with only 8 examples previously described in the English literature. Author described adenocarcinoma emerging from a recurring adult teratoma in the right frontal convexity outside of the pineal region, and genetic examination of the recurrent tumour revealed a mutation in KRAS. Somatic-type carcinomas are a potential diagnostic stumbling block because most pineal germ cell tumours are diagnosed by biopsy rather than extensive tissue sampling [4].

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First, a mucin-producing carcinomatous component can be mistaken for metastatic carcinoma. Second, its absence in the biopsy specimen does not rule out its presence in the tumor's unsampled areas. The expression of the transcription factor CDX2, which is often utilised as a marker of lower intestine differentiation/origin, is not helpful in separating somatic-type adenocarcinoma from yolk sac tumour, as shown in the current case. In patients with intracranial yolk sac tumours, malignant somatic transformation is an uncommon cause of recurrent tumours or treatment failure. Neoadjuvant chemotherapy and craniospinal irradiation are the most common treatments for yolk sac cancers [5]. Surgery is usually reserved for individuals who have failed to respond to other treatments. However, given the known poor response of patients with extracranial nongerminomatous germ cell tumours to cisplatin-based chemotherapy, expanding the existing treatment paradigm to include resection as a first-line therapy may be worth considering and warrants additional exploration.

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