Disseminated hematogenous metastases from a pineal germinoma in an infant

Case report

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A pineal germinoma in a 9-month-old boy is described. After surgical resection and cerebrospinal fluid
diversionary shunting, the child developed hematogenous metastases which showed marked increased uptake
on radionuclide gallium-67 scans.

KEY WORDS • pineal tumor • germinoma • hematogenous metastasis • gallium-67

Tumors of the pineal region constitute 2.7% of all intracranial neoplasms and, among them, pinealoma (including germinoma) has the highest incidence in the series of Sano, et al.12 The Brain Tumour Registry in Japan reports that germinomas comprised 2.4% of 6204 cases of primary intracranial neoplasms.13 This tumor occurs most frequently in patients aged between 10 and 15 years. Hematogenous metastases from an intracranial germinoma is a rare occurrence, usually found after intracranial surgery or cerebrospinal fluid (CSF) diversionary shunting.4,7,11 In the evaluation of tumor recurrence and metastases, gallium-67 citrate (67Ga) is being increasingly utilized, in both adults and children.1,5 The application of 67Ga has not been reported in the detection of germinomas.

Case Report

This 9-month-old boy presented with a 5-week history of a squint and an inability to push himself around in his “walker,” although he had done so previously. Two weeks prior to admission, his parents noted a right hemiparesis, and the child became very irritable and began vomiting frequently.

Examination. The patient was drowsy. He had a large head (47.5 cm, greater than the 97% percentile) and a tense fontanel. His eyes had the “setting sun” appearance, and his pupils were equal but poorly reactive to light. There was no papilledema. Reflexes were generally brisk, and he had a positive Babinski sign bilaterally and a right hemiparesis. A computerized tomography (CT) scan (Fig. 1) revealed a large enhancing tumor at the midline, arising from the pineal region. There appeared to be cystic extension into the left lateral ventricle association with a marked obstructive hydrocephalus.

Operations. At surgery via a left parieto-occipital craniotomy and a transcortical transventricular approach, decompression of the cystic portion and sub-

![Fig. 1. Computerized tomography scans. Left: Plain scan showing a large midline tumor with cystic extension into the left lateral ventricle associated with markedly dilated ventricles. Right: After contrast injection the midline solid component of the pineal tumor and rim of the cystic component enhance markedly.](image-url)
total resection of the solid tumor was carried out. Histology confirmed the diagnosis as a germinoma. Postoperatively, the child was more alert, but developed Parinaud's syndrome. Repeat CT scanning showed a large amount of necrotic tissue and moderate bilateral subdural collections. The patient's condition deteriorated and bilateral subdural-peritoneal shunts were inserted. One month later, a further subtotal resection was carried out via a supratentorial, transcallosal approach. Following operation obstructive hydrocephalus occurred. Bilateral ventriculoperitoneal shunts were inserted and connected to the subdural-peritoneal shunt systems. The patient improved and underwent 5 weeks of craniospinal irradiation. Repeat CT scans showed marked reduction in tumor size over this period.

Postoperative Course. After discharge, the child progressed well for 9 months after his initial presentation. At this time, he developed a painful left hip. On examination a firm mass was palpable in the upper lateral portion of his left thigh. An x-ray film revealed areas of bone erosion in the left upper femur and ischium. A bone scan with technetium-99m methylene diphosphonate revealed multiple focal areas of increased uptake in the proximal left femur, ischium, ribs, ilium, and in the skull, indicating multiple skeletal metastases. A $^{67}$Ga citrate study showed avid accumulation of tracer in the soft tissues of the left thigh, skull (particularly the frontal cranial region), ribs, femora, groin, and abdomen (Fig. 2). The chest x-ray was normal. The child was commenced on chemotherapy comprising cisplatinum, bleomycin, and vinblastine. The patient died 2 months later with no significant regression of his metastases. Permission for an autopsy was not obtained.

Discussion

Intracranial germinoma with hematogenous metastases is a very rare occurrence in childhood. A child of 9 months with pineal germinoma has not been reported in the literature; the usual age at presentation being from 10 to 25 years, especially between 10 and 15 years. Germinomas usually invade locally into the ventricular system and then by seeding via the subarachnoid space. Extradural spread may occur rarely with invasion of the petrous bone, carotid artery, and other adjacent structures. Dissemination into the CSF space and via diversionary shunts has been reported to occur in 8% to 20% of patients. Haimovic, et al., discussed a case of germinoma with large abdominal metastases secondary to seeding via the diversionary shunt. In five reported cases of hematogenous pulmonary metastases, spread was detected in each case after subtotal excision of the primary pineal tumour. Borden, et al., reported a case of a 9½-year-old boy who presented 1 year after diversionary CSF shunting and radiotherapy with bone metastases in the femoral neck associated with a large soft-tissue tumor. This child subsequently had multiple metastases to the

Fig. 2. Left: Gallium-67 citrate ($^{67}$Ga) whole-body scan demonstrating multiple skeletal and soft-tissue metastases. There is marked avid accumulation of tracer in the frontal skull, right maxilla, right proximal humerus, and left ribs (arrows). Center: Posterior view of the trunk showing $^{67}$Ga uptake in the ribs, pelvis, and left thigh. Right: Lower limbs showing increased uptake of $^{67}$Ga in the left thigh, distal right femur, and proximal right tibia.
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bone, lymph nodes, and pleura; however, he responded to chemotherapy and was alive 11 years later.

The established therapy of pineal germinoma is by direct surgery and postoperative irradiation. Germi-
noma patients under 15 years old showed better survival rates than those over 15 years. The greatest criticism against direct surgery, however, is the increased prob-
ability of dissemination of metastases in the CSF space.

Bone scanning with technetium-99m phosphate compounds has been shown to be the most sensitive method for detecting bone metastases in both children and adults. Gallium-67 citrate is also valuable in investigating pediatric solid tumors that are initially Ga-avid, and has been effectively used for detection of recurrence and metastases. Positive Ga accumulation has been reported in seminomas but no reference in the literature was found for germinomas. Gallium-67 scans should be performed in the initial staging of patients with germinoma to determine whether the tumor is gallium-avid, and to detect metastatic disease. In those patients where this is confirmed then, serial Ga scans at 6-monthly intervals or as the clinical situation determines would be useful to assess the response to therapy and to diagnose early recurrence or further metastatic spread.

References


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