Anaplastic Astrocytoma of an Oncocytic Type Occurring in the Cerebellar Vermis in Pierre Robin Syndrome

—Case Report—

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Abstract

A 44-year-old male with Pierre Robin syndrome and funnel chest was diagnosed with a tumor of the cerebellar vermis and spontaneous pneumothorax. He received tube thoracostomy for pneumothorax and ventriculoperitoneal shunt for hydrocephalus, followed by radiological examination, subtotal removal of the tumor, pneumonorrhaphy, and chemoradiotherapy. Light microscopy of the tumor sample revealed marked pleomorphism of the tumor cells and numerous giant cells, without mitotic figures. Microcystic changes due to vasogenic edema were also evident throughout the tumor. Electron microscopy showed more than 50% of all tumor cells to be oncocytes, with numerous mitochondria in the cytoplasm. The tumor was diagnosed as anaplastic astrocytoma of an oncocytic type.

Key words: anaplastic astrocytoma, oncocytic transformation, Pierre Robin syndrome, funnel chest, pneumothorax

Introduction

Pierre Robin syndrome is a congenital malformation characterized by micrognathia, glossoptosis, and cleft palate.¹³⁾ We describe an adult male with this condition who developed cerebellar anaplastic astrocytoma of an oncocytic type, and report the many difficulties in pre- and postoperative managements.

Case Report

A 44-year-old male complained of headache and vomiting at the end of February, 1988. He was admitted to our hospital on March 12. His history included conservative treatment for spontaneous pneumothorax at another hospital in April, 1987. His family history was non-contributory.

Physical examination on admission showed micrognathia, glossoptosis, cleft palate, and funnel chest. Neurological abnormalities included in-

tracranial hypertension and truncal ataxia. Plain skull x-ray films disclosed micrognathia (a typical sign of Pierre Robin syndrome), but no signs of intracranial hypertension (Fig. 1). Plain chest x-ray

Fig. 1 Plain skull x-ray film, disclosing micrognathia.

Fig. 2 *left*: Precontrast CT scan, disclosing an isodense mass on the midline in the posterior fossa. *right*: Postcontrast CT scan, showing the heterogeneously enhanced mass.

Fig. 3 MR imaging, showing the tumor as low intensity on the T₁-weighted image (*left*) and high intensity on the T₂-weighted image (*right*).

films revealed incidental pneumothorax on the right. Tube thoracostomy was therefore performed to start continuous low-pressure aspiration.

Precontrast computed tomographic (CT) scans disclosed an isodense mass on the midline in the posterior fossa (Fig. 2 *left*). Postcontrast CT scans showed heterogeneous enhancement (Fig. 2 *right*). Based on these findings and the presence of severe hydrocephalus, a ventriculoperitoneal (VP) shunt was emplaced on the first hospital day. During this operation, intubation for general anesthesia was difficult using routine methods because of Pierre Robin syndrome. Therefore, intubation was performed using a bronchoscope. When the endotracheal tube was withdrawn at the end of the operation, he developed marked cyanosis due to depression of the tongue, which necessitated re-intubation and a tracheostomy.

Magnetic resonance (MR) imaging disclosed a solid tumor in the cerebellar vermis, which was low

intensity on T₁-weighted images (Fig. 3 *left*) and high intensity on T₂-weighted images (Fig. 3 *right*). Cerebral angiograms revealed tumor stains supplied by the bilateral posterior inferior cerebellar and bilateral superior cerebellar arteries. A suboccipital craniectomy on April 14 removed the hemorrhagic cerebellar tumor subtotally. Pneumonorrhaphy for spontaneous pneumothorax was undertaken on May 6. Subsequently, he received radiation therapy (30 Gy whole brain, 40 Gy local) with ACNU and vincristine.

He was discharged on August 31 although truncal ataxia persisted. He received maintenance therapy using ACNU and procarbazine at our outpatient clinic. On July 27, 1989, he died due to tumor recurrence. His family refused an autopsy.

The surgical specimen was examined. Light microscopy revealed marked pleomorphism of the tumor cells, appearance of giant cells, and vascular

Fig. 4 Histological examination of the surgical specimen. *upper*: Light microscopy, revealing marked pleomorphism of the tumor cells, and microcystic changes due to vasogenic edema. HE stain, $\times 100$. *lower*: Electron microscopy, showing numerous oncocytes in the cytoplasm and enlargement of extracellular spaces due to vasogenic edema. Bar = 1 μ m.

proliferation in some places, although mitotic figures were absent (Fig. 4 upper). Microcystic changes, suggestive of vasogenic edema, were evident throughout the tumor. No necrotic area was visible. Immunohistochemical examination showed positive staining for glial fibrillary acidic protein and S-100 protein. The mean number of argyrophilic nucleolar organizer regions was 2.7, suggesting high-grade glioma. Electron microscopy showed more than 50% of tumor cells to be oncocytes possessing numerous mitochondria in the cytoplasm (Fig. 4 lower). The extracellular spaces were enlarged by vasogenic edema. Based on these findings, the tumor was finally diagnosed as anaplastic astrocytoma of an oncocytic type.

Discussion

Hamperl¹⁴⁾ first introduced the term "oncocyte" into pathology in 1931. It originates from a Greek word "ὄγκος (onkos)" which means mass or bulk. An oncocyte is more than double the size of a normal cell. The cytoplasm contains many eosinophilic granules. Oncocytes are also observed in normal tissues such as the salivary glands, thyroids, and parathyroids,⁵⁾ and increase in numbers with age.¹⁵⁾ Since the cytoplasm of oncocytes contains numerous mitochondria, these cells are thought to represent a mitochondrial disorder of epithelial cells.¹⁵⁾ In 1932, Jaffé⁶⁾ used the term "oncocytoma" to indicate an oncocyte-rich parotid gland tumor. At present, the term "oncocytoma" is designated for use where more than 50% of all tumor cells are oncocytes.¹²⁾

Oncocytic transformation of primary brain tumors has occurred in pituitary adenoma,^{9,11)} anaplastic glioma,^{3,7,8,10)} and choroid plexus papilloma.^{1,14)} All investigators regarded it as a degenerating process. The present case is the first reported case of anaplastic astrocytoma of an oncocytic type in the posterior fossa. The relationship between the oncocytic transformation and prognosis should be studied.

Pierre Robin syndrome is a congenital malformation characterized by micrognathia, glossoptosis, and cleft palate. This syndrome is frequently complicated by congenital heart disease, thoracic hypoplasia, and glaucoma.¹³⁾ Intracranial complications, such as microcephaly and hypoplasia of the cerebellar vermis, have also been reported.^{2,13)} However, primary brain tumor has not previously occurred as complication of Pierre Robin syndrome. The present case was complicated by funnel chest and spontaneous pneumothorax in addition to the

brain tumor.

The initial intention was treatment with tube thoracostomy and VP shunt, followed by precise neurological examination, removal of the brain tumor, pneumonorrhaphy, and subsequent chemoradiotherapy. This original plan, however, had to be modified because tracheostomy was required just after the VP shunt. As the respiratory function in such cases may be restricted by Pierre Robin syndrome and funnel chest, we recommend tracheostomy prior to the VP shunt.

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