

M. Nagayama<sup>a</sup>  
 Y. Shinohara<sup>a</sup>  
 T. Takeoka<sup>a</sup>  
 M. Haida<sup>a</sup>  
 A. Ikeda<sup>b</sup>  
 I. Yamamoto<sup>b</sup>  
 O. Sato<sup>b</sup>

Departments of

<sup>a</sup> Neurology and

<sup>b</sup> Neurosurgery, Tokai University, School of  
 Medicine, Isehara, Kanagawa, Japan

# Intracranial Paramedian Hourglass-Shaped Dermoid Associated with Hereditary Steatocystoma multiplex

## Key Words

Dermoid  
 Steatocystoma multiplex  
 Hemifacial spasm  
 Magnetic resonance imaging

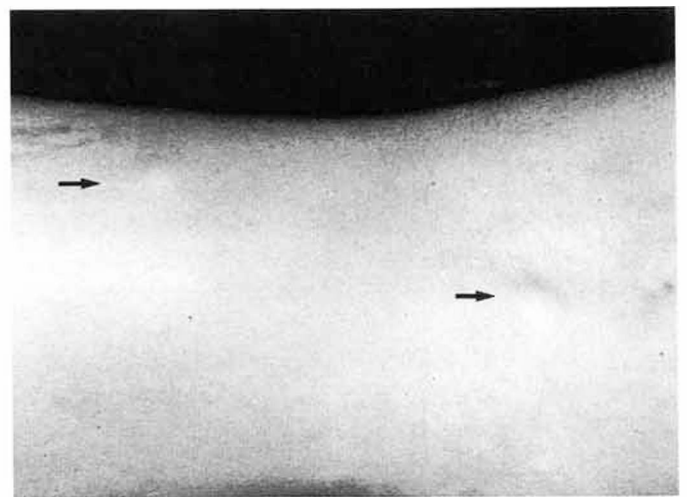
## Abstract

This is the first report to describe the coexistence of two rare diseases, intracranial paramedian hourglass-shaped dermoid and steatocystoma multiplex. A 46-year-old female with a history of steatocystoma multiplex, bradydactyly and kyphosis showed oscillopsia, ataxia and hemifacial spasm. MRI findings suggested a giant dermoid cyst extending from the left middle temporal fossa to the cerebellopontine angle, and this was confirmed surgically. We propose the involvement of some genetic factor or pathological process common to both disorders, steatocystoma multiplex and dermoid. From the clinical point of view, patients with cystic skin lesion should also be checked for intracranial lesion.

Neurocutaneous syndrome includes heterogeneous disorders linked only by the common property of involvements of the skin and nervous system, both of which are of ectodermal origin [1-3]. The purpose of this communication is to report the first patient with intracranial paramedian hourglass-shaped giant dermoid associated with steatocystoma multiplex, a rare retention cyst of the follicular and sebaceous system, and to propose a possible common etiology for these two disorders.

## Patient Report

A 46-year-old female had noted a subcutaneous mass at her buttocks at first in her 30s. Thereafter, the masses gradually spread over the whole body, but she did not seek medical advice until 1986 at the age of 46, when she noted clumsiness of the left upper extremity, oscillopsia, dizziness, and numbness of the left cheek. She was



**Fig. 1.** Multiple subcutaneous cystic masses (arrows), confirmed histopathologically as steatocystoma multiplex.

Received:  
 April 24, 1991  
 Accepted:  
 August 28, 1991

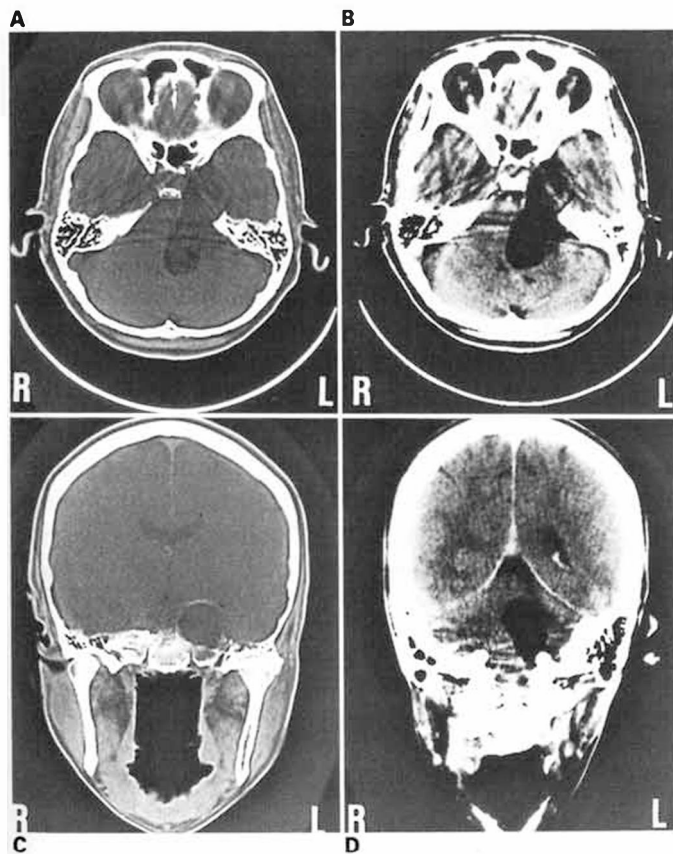
M. Nagayama, MD  
 Department of Neurology  
 Tokai University, School of Medicine  
 Isehara, Kanagawa 259-11 (Japan)

© 1992 S. Karger AG, Basel  
 0014-3022/92/0325-0260  
 \$2.75/0

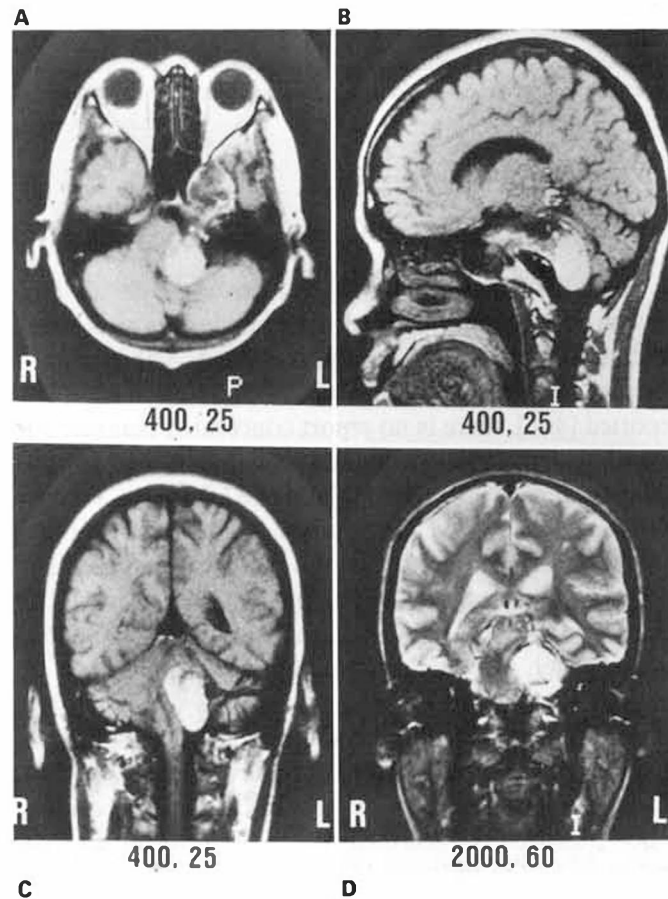
referred to our hospital in January 1987. Her history included surgery for flexion contracture of right toe and hypertension. Her mother had the same subcutaneous lesions. Intermarriage was not noted. Physical findings were as follows. Height 160 cm, weight 51 kg, pulse 74/min and blood pressure 144/80 mm Hg. Multiple subcutaneous cystic masses were observed over the whole body, especially at both upper extremities. The masses were approximately 5–20 mm in diameter (fig. 1). Bradydactylies and kyphosis of the spine were also noted. Neurologically, consciousness, mental state and speech were normal. Papilledema was present bilaterally. Saccadic eye movements and Bruns-Cushing nystagmus with fine rapid movements to the right were observed. Paresthesia at the territory of the ophthalmic branch of the trigeminal nerve, decreased corneal reflex, lower eyelid spasm, and hearing loss were noted on the left. Gag reflexes were decreased bilaterally. Motor and sensory systems were otherwise normal. Reflexes were normal except for positive snout reflex. Cerebellar signs were observed in the left side. Romberg's sign was negative. Meningeal signs were also absent. Laboratory data were normal except for iron deficiency anemia. Chromosomal examination was normal.

Skull X-P showed deformed bilateral internal acoustic meatus and osteolytic changes of the sphenoid sinus and of the left petrosal bone. Computed tomography (CT) (fig. 2) disclosed a paramedian hourglass-shaped giant cystic mass, extending from the left middle temporal fossa to the left cerebellopontine angle, associated with destruction of the petrous apex as shown on the left side of figure 2A, C. Linear marginal enhancement was also apparent. Magnetic resonance imaging (MRI) (fig. 3) showed the known giant cystic mass. The T1 weighted images (TR/TE 400/25 ms) in sagittal view disclosed the anterior part of the mass to be of low intensity, and the posterior part of high intensity, with an irregularly shaped boundary. So it was suspected that the posterior part of the mass consisted of short T1 material such as fat tissue. Angiography (left CAG and right VAG) documented the presence of a hypovascular mass suggesting extramedullary location.

From these findings, the intracranial mass was considered as epidermoid or dermoid. In April 1987 a surgical procedure was performed. Some hair was observed in the tumor macroscopically, and microscopic findings showed that the tumor capsule consisted of keratin, and the cystic fluid contained cellular keratin debris. On the basis of these pathological findings we diagnosed this as dermoid.



**Fig. 2.** Plain CT showed a paramedian hourglass-shaped giant cystic mass (B, D), associated with bony destruction (A, C).



**Fig. 3. A–D** MRI. T1 weighted images (TR 400 ms, TE 25 ms) showed the anterior part of the mass consist of long T1 material such as water, and the posterior part of short T1 material such as fat.



**Fig. 4.** Microscopic finding of the subcutaneous mass. Sebaceous gland in the follicular epithelial wall, which surrounds the cavity, provides the diagnosis of steatocystoma multiplex. H-E.  $\times 360$ .

Microscopic examination of the subcutaneous cystic mass indicated steatocystoma multiplex (fig. 4), because sebaceous glands were observed in the follicular epithelial wall which surrounded the cystic cavity.

## Discussion

The following three clinical features of the dermoid in this patient are noteworthy. First, it is giant in size, paramedian in location and hourglass-shaped. Although rare cases of paramedian hourglass epidermoid have been reported [4–6], there is no report concerning paramedian hourglass dermoid as far as we know. The second feature is the discrepancy between CT and MRI findings. Though CT images suggested the entirely cystic nature of the mass, the MRI indicated that the mass was composed of at least two heterogeneous components. MRI showed that the anterior part of the mass consisted of long T1 material such as water, and the posterior part consisted of short T1 material such as fat. From these findings we tentatively assumed this mass to be dermoid or epidermoid preoperatively. As is well known, the contents of dermoid consist of a mixture of lipid (low density in CT), keratin and its debris (iso-density), secreted materials from the capsule and so on. So the ability of MRI to discriminate the heterogeneity of dermoid is a great advantage, especially in cases with high lipid content, in which CT shows only a diffuse low density region [7]. The third feature is the presentation of hemifacial spasm. Although cases of intra-

cranial epidermoid with hemifacial spasm have been reported [8–14], we could not find this association for dermoid in the literature. The hemifacial spasm in this patient may be due to extra-axial compression of the facial nerve roots by the tumor.

On the other hand, steatocystoma multiplex is a very rare retention cyst of the follicular and sebaceous systems, often inherited as an autosomal dominant trait, and characterized by numerous intradermal cysts scattered over the whole body [1–3, 15]. The cystic wall is composed of stratified squamous epithelium, and the presence of sebaceous glands in the wall is crucial. The contents are composed of pure lipid including cholesterol. The lipid contents are fairly saturated, approximately half-way between blood lipid and depot fat. Hairs may also be found in the contents. Two main theories of the pathogenesis have been proposed [16]. The retention theory is that obstruction of the sebaceous excretion duct due to keratinization causes dilation and cystic formation of the pilosebaceous junction. The other theory is that, because of the coexistence of hairs, hair follicles, sebaceous and eccrine glands, the tumor is pluripotential in nature, and is a kind of dermoid tumor or hamartoma histopathologically.

In our case, the clinical and histopathological findings, including family history, were characteristic of this disease. The most important point was whether the association of the two diseases observed in this patient is coincidental. Both disorders are congenital epithelial tumors, and dermoid contains hairs, hair follicles, sebaceous glands and eccrine glands, as does steatocystoma multiplex. Therefore pathological findings of both disorders are quite similar. So we would like to propose the involvement of some genetic factor or pathological process common to both disorders. From the clinical point of view, clinicians should take care to examine whether patients may have intracranial lesion as well as cystic skin lesion.

---

## References

- 1 Adam RD: Neurocutaneous diseases; in Fitzpatrick TB, Eisen A, Wolff K, Freedberg IM, Austen KF (eds): *Dermatology in General Medicine*. New York, McGraw-Hill, 1987, pp 2022–2025.
- 2 Mackic RM: Tumors of the skin; in Rook A, Wilkinson DS, Ebling FJG, Champion RH (eds): *Textbook of Dermatology*, ed. 4, Oxford, Blackwell, 1986, vol 2, pp 2406–2407.
- 3 Caro WA, Bronstein BR: Tumors of the skin; in Moschella SL, Hurley HJ (eds): *Dermatology*, ed 2. Philadelphia, Saunders, 1985, vol 2, pp 1541–1542.
- 4 Albanese V, Tomasello F, Picozzi P, Sampaolo S, Spadaro A: Paramedian hourglass epidermoid extending above and below the tentorium. *Surg Neurol* 1982;18:356–363.
- 5 Imamura Y, Ninchoji T, Nakajima S, Uemura K: Epidermoid tumor in the fourth ventricle, with particular reference to metrizamide CT cisternography findings. *Surg Neurol* 1982;18:444–447.
- 6 Ogawa T, Sekino H, Fuse T, Nakamura N: Multiple intracranial epidermoids located in the brain stem and the middle cranial fossa. *Neurol Med Chir (Tokyo)* 1985;25:393–397.
- 7 Horowitz BL, Chari MV, James R, Bryan RN: MR of intracranial epidermoid tumors: Correlation of in vivo imaging with in vitro  $^{13}\text{C}$  spectroscopy. *AJNR* 1990;11:299–302.
- 8 Auger R, Piepgras D: Hemifacial spasm associated with epidermoid tumors of the cerebellopontine angle. *Neurology* 1989;39:577–580.
- 9 Otsuka S, Nakatsu S, Matsumoto S, Sato S, Motozaki T, Ban S, Yamamoto T, Shirane H: Epidermoid tumor presenting with trigeminal neuralgia and ipsilateral hemifacial spasm: A case report. *Arch Jpn Chir* 1989;58:245–249.
- 10 Sprik C, Wirtschafter JD: Hemifacial spasm due to intracranial tumor: An international survey of botulinum toxin investigators. *Ophthalmology* 1988;95:1042–1045.
- 11 Miyazaki S, Fukushima T: CP angle epidermoid presenting as hemifacial spasm. *No To Shinkei* 1983;35:951–955.
- 12 Liebaltd GP, Henn R: Vergleichende klinisch-anatomische Gesichtspunkte beim 'spasmus facialis' bei einem Fall von Epidermoid der hinteren Schädelgrube. *Dtsch Z Nervenheilk* 1970;197:149–162.
- 13 Arseni C, Petrovici I: Persistent tonic facial spasm in brain stem tumors. *J Neurol Sci* 1968;7:107–114.
- 14 Revilla AG: Differential diagnosis of tumors at the cerebellopontine recess. *Bull Johns Hopk Hosp* 1948;83:187–212.
- 15 Ormsby OS, Finnerud CW: Steatocystoma multiplex. *Arch Dermatol Syphilol* 1930;22:822–827.
- 16 Kligman AM, Kirschbaum JD: Steatocystoma multiplex: A dermoid tumor. *J Invest Dermatol* 1964;42:383–387.