Original

Pneumosinus Dilatans Associated with Orbital Neurilemmoma

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Key Words
orbit;
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pneumosinus;
schwannoma

Backgroound. *Pneumosinus dilatans (PSD)* is a rare condition. It has been reported in association with spheno-orbital meningioma, optic nerve sheath meningioma, orbital fibro-osseous diseases and intracranial meningioma, but never with orbital neurilemmoma. We report 4 cases of orbital neurilemmomas in association with PSD, and stress the importance of PSD in cases of orbital neurilemmomas.

Methods. We reviewed the hospital chart, and found 324 orbital tumors were found from 1974 to 1996. Eight consecutive cases of pathology-proven primary orbital neurilemmoma were studied. Clinical features, especially computed tomography (CT) scan, were completely reviewed to find the co- existence of orbital neurilemmoma and pneumosinus dilatans.

Results. Four cases (50%) among 8 cases of pathology-proven orbital neurilemmomas were found to have pneumosinus dilatans in their CT scan. All these four cases were extraconal tumors. Representative CT scan showed enlarged, aerated sinus with thinning of the affected sinus walls. Ethmoidal, frontal and maxillary sinuses were the most frequently affected.

Conclusions. *Pneumosinus dilatans* is commonly associated with primary orbital neurilemmoma. [*Chin Med J* (*Taipei*) *2002;65:218-224*]

Four percent of orbital neoplasms are periperal nerve tumors and primarily consist of neurofibromas and neurilemmomas (or schwannomas). Of these, 2% of the lesions are plexiform neurofibromas, 1% isolated neurofibromas, and 1% neurilemmomas (or schwannomas).[1,2] Thus, neurilemmoma is a relatively uncommon tumor in the orbit. This benign Schwann cell tumor of peripheral nerves usually presents as a slow-growing tumor in adulthood, with the age at diagnosis ranging from 20 to 70 years.[3] There is no predilection for sex, but some reported that women are slightly more commonly affected.[3,4] Most cases are unilateral and solitary. Ocular proptosis, decreased vision, diplopia, trigeminal distribution numbness or pain occurred as the presenting clinical features.[4] The direction of proptosis may vary, but usually with downward displacement of the globe, as most neurilemmomas involve the superior orbit and arise from branches of either the supraorbital or supratrochlear nerve.[3,4] Orbital neurilemmoma is an encapsulated, noninvasive lesion that has minimal effects on other orbital structures except by mechanical compression.[5] On computed tomography (CT), it shows a well-defined homogenous mass that enhances mildly to moderately.[4,5] Calcification within the tumor is rare, but cystic change is characteristic. Meanwhile, bony expansion with enlargement of superior or inferior orbital fissures is not uncommon.[4] Ultrasonography usually shows a solid lesion with a sharply outlined capsule. The tumor may contain a central cystic space due to myxoid degener a-

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tion.[5,6] On magnetic resonance imaging (MRI), a more heterogenous appearance is not uncommon. On T2-weighted scans, these tumors are typically hyperintense to brain and isointense with cerebrospinal fluid.[4]

Pneumosinus dilatans (PSD) is a rare condition. It refers to an abnormal dilatation of one or more paranasal sinuses without radiologic evidence of localized bone destruction, hyperostosis or mucous membrane thickening.[7,8] It may affect any of paranasal sinuses and most often affects the frontal sinus.[8] PSD has been reported to be in association with suprasellar meningioma,[9] optic nerve sheath meningioma of optic canal,[10-12] orbital fibro-osseous diseases[13-16] and intracranial meningioma involving the frontal lobe.[17] To our knowledge, PSD associated with orbital neurilemmoma has never been reported previously. Here we report 4 cases of orbital neurilemmomas in association with PSD.

Methods

We conducted a retrospective review of orbital tumors diagnosed at Taipei Veterans General Hospital. The computer registry of Taipei Veterans General Hospital was searched for the diagnosis of orbital tumor, and a total of 324 patients were found to have orbital tumors from 1974 to 1996. Among these patients, 8 (2.4%) cases of pathology-proven primary orbital neurilemmomas were found. We analyzed the clinical features, especially the CT scan, in these patients. The diagnosis of PSD was based on standard radiography, CT scan or MR imaging study. The diagnostic criteria for PSD consists of (1) the enlargement of an air cell or the whole sinus, (2) the presence of only air in the abnormal space, and (3) the ballooning outward of the walls of the sinus, which may be thinned and/or demineralized.[18]

Results

From 1974 to 1996, eight cases of pathology-proven orbital neurilemmomas were collected, including 6 males and 2 females aged from 24 to 69 years (mean age of 44.2 years). Five patients presented with orbital mass in the right orbit, while 3 in the left orbit. Tumors were located extraconally in 6 and intraconally in 2.

Clinical symptoms and signs revealed proptosis (8/8), EOM limitation (6/8), visual impairment (3/8) and lid swelling (2/8). Four patients were found to have pneumosinus dilatans in their CT scan. In these 4 cases, the orbital masses were all located extraconally.

Case 1

A 24-year-old male patient presented with progressive proptosis of right eye for 5 months. Vision was 6/4.5 in both eyes. Limitation of upper gaze was noted in the right eye. Pupillary light reflex was prompt in both eyes. Exophthalmometer showed 6 mm proptosis of the right eye in comparison with left eye. Ophthalmoloscopy revealed choroidal fold in the right eye. CT scans (Fig. 1 ABC) showed a large orbital mass with marginal enhancement, located superiorly and posteriorly in orbital apex. Pneumosinus dilatans of frontal, sphenoidal, ethmoidal and maxillary sinuses were noted as well. The right orbital mass was removed subtotally through lateral orbitotomy. Histopathological examination revealed a typical picture of neurilemmoma (Fig. 1D) consisting of Antoni-A, Antoni-B cells, Verocay bodies and positive S-100 protein stain. Postoperative course was smooth. Vision of both eyes remained 6/6 with free movement of both eyes during a 16-month follow-up period.

Case 2

A 32-year-old female had progressive protrusion of left eye and occasional diplopia for 1 year. Her best corrected vision was 6/4.5 in the right eye and 6/7.5 in the left eye. Slitlamp biomicroscopy revealed normal anterior segment. Horizontal retinochoroidal folds was found in the left eye with fundoscopy. Exophthalmometer showed 5 mm proptosis of the left eye compared with right eye. CT scans showed an extraconal orbital mass located in superior and lateral part of left oribit, with thinning of the left orbital roof. PSD of sphenoid, ethmoidal and frontal sinuses were also noted (Fig. 2). Lateral orbitotomy with complete

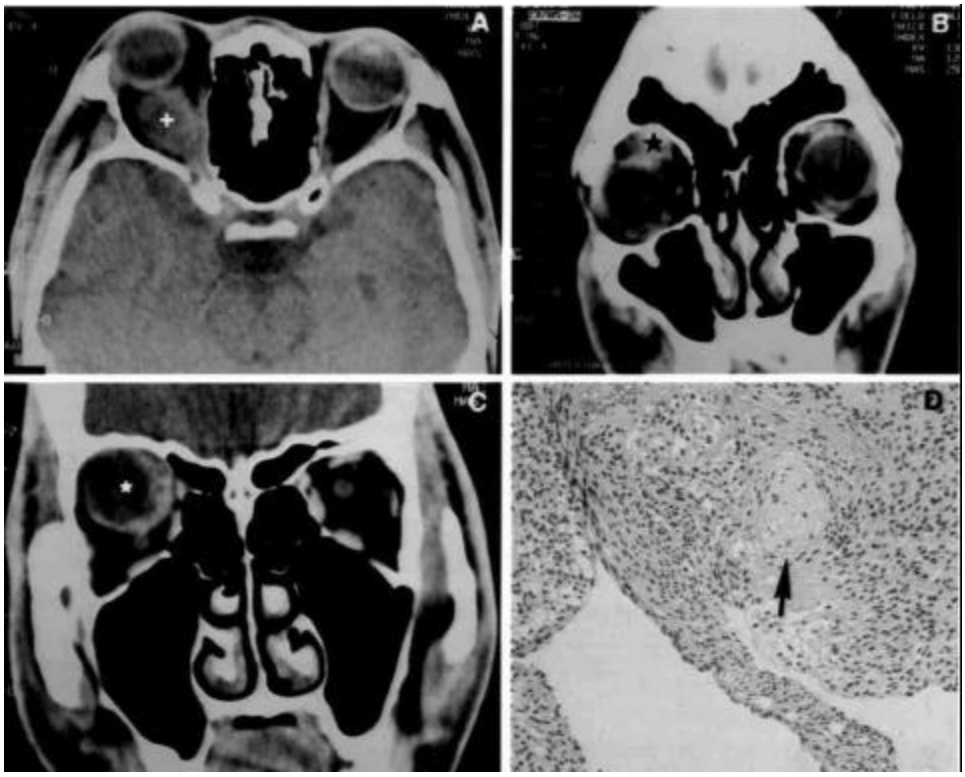

Fig. 1. Clinical and pathological profiles of case 1. (**A**) Axial orbital mass with heterogenous intensity (+) located in retrobulbar space of orbit; (**B**) The tumor (*) in the superior orbit displaced the right eye downward; Pneumosinus dilatans of the frontal and ethmoidal sinuses were also noted; (**C**) Marginal enhancement on the nasal periphery of the tumor; and (**D**) Verocay body within the tumor (arrow). (hematoxylin-eosin, × 40).

removal of orbital tumor was performed. Histopathological examination disclosed a neurilemmoma with myxomatous change. Postoperative course was smooth and the patient was well during a 4-year follow-up period.

Case 3

A 60-year-old male complained of decreased vision in his right eye and diplopia for one and a half years. Best corrected visual acuity was 6/6 in both eyes. Exophthalmometer showed 5 mm proptosis of the right eye compared with left eye. Slitlamp biomicroscopy revealed a pterygium in the right eye. Mild limitation of adduction was noted in the right eye. CT scans demonstrated an extraconal orbital mass in the medial orbit, and PSD of ethmoidal and sphenoidal sinuses (Fig. 3A and Fig. 3B). Anterior orbitotomy was performed to remove the tumor. Histopathological examination disclosed a grayish soft tumor with size of $1.2 \times 1.2 \times 2$ centimeter. Microscopically, the section showed a picture of neurilemmoma composed of cellular mass with elongated nuclei in a whorled pattern in a collagenous matrix. Palisading of nuclei is conspicuous. The patient was rather well with corrected vision of 6/10 in the right eye and 6/6 in the left eye after 4 years of follow-up.

Case 4

A 27-year-old male presented with right eye protrusion and visual impairment for more than 2 years. His visual acuity was 6/10 in the right eye and 6/6 in the left eye. PSD of ethmoidal and sphenoidal sinuses were seen on CT scan (Fig. 3C and Fig. 3D). The right orbital tumor was completely removed. Histopathological examination proved to be neurilemmoma. No signs of tumor recurrence were noted during a 2-year follow-up period.

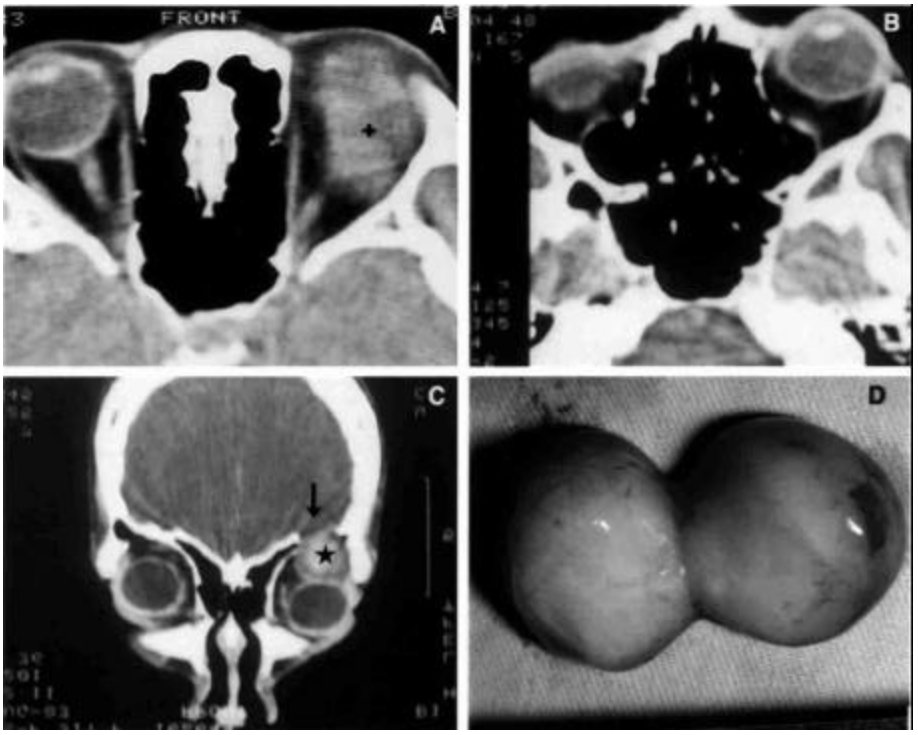

Fig. 2. Clin i cal and patho log i cal pro files of case 2. (**A**) an or bital mass with heterogenous in ten sity (+) was lo cated in su perior part of left or bit; (**B**) Pneumosinus dilatans of the sphenoidal si nus; (**C**) The tu mor (*) in the su pe rior or bit displaced the left eye down ward; Cen tral en hance ment of the tu mor and bony ero sion of the left o r bital roof were found (ar row); and (**D**) Gross ap pear ance and cut sur face of the re moved tu mor spec i men.

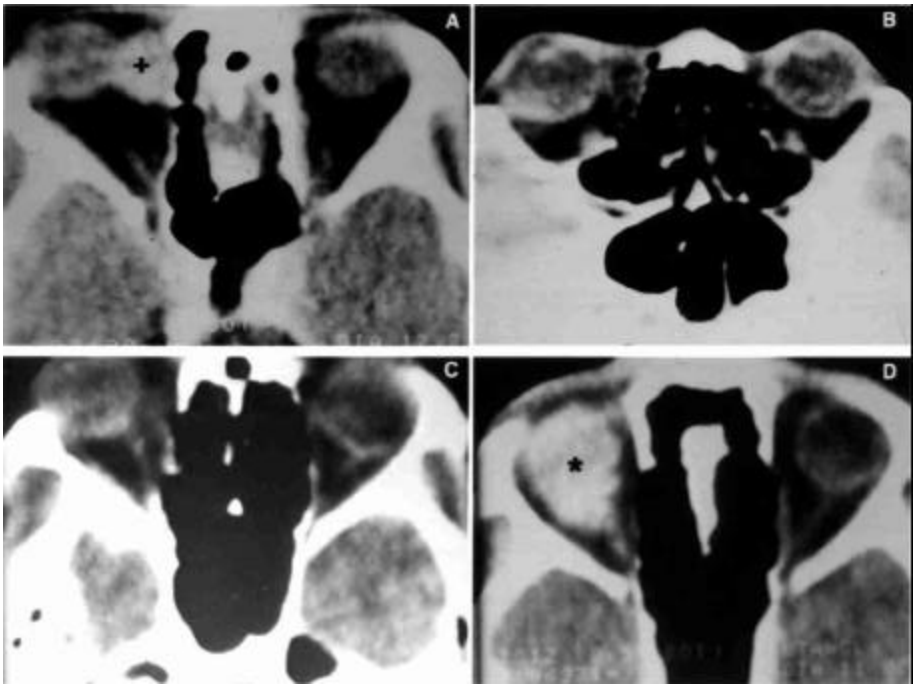

Fig. 3. CT scans of case 3 and case 4. (**A**) An or bital tu mor lo cated at the me dial or bit (+); (**B**) Pneumosinus dilatans of the sphenoidal si nus; (**C**) Pneumosinus dilatans of the ethmoidal si nus and an te rior clinoid pro cess; and (**D**) A well enhanced tu mor (*) in the right or bit.

Discussion

Neurilemmoma, or schwannoma, a peripheral nerve sheath tumor, is a pure proliferation of Schwann cells. This Schwann cell tumor usually presents as a slowly progressive tumor in the adulthood. It has been reported to associate with neurofibromatosis type 1. Approximately 1.5% to 18% of patients who have neurofibromatosis have orbital neurilemmoma.[19] Malignant transformation of the neurilemmoma has been known to occur in patients with neurofibromatosis,[19] though malignant transformation is extremely rare. Histopathologically a benign neurilemmoma is well differentiated and is encapsulated in the perineurium of the nerve of its origin. The classic picture is the alternation within the tumor of solid cellular area, referred to as the Antoni A pattern, and loose myxoid tissue with stellate or ovoid nuclei, referred to as an Antoni B pattern.[20] Cystic change within the tumor may occur secondary to coalescence of mucinous or microcystic areas in Antoni B tissue of neurilemmomas.[3]

The echographic finding of neurilemmomas shows a solid lesion with a sharply outlined capsule. The tumor may contain a central cystic space due to myxoid degeneration.[3,5,6] The internal reflectivity was low within the cystic spaces, and was medium to high within the surrounding solid regions.[6] On CT scan, neurilemmomas show a well-defined homogenous mass that enhances mildly to moderately.[4,5] Calcification within the tumor is rare, but cystic change is characteristic. CT scan is the most useful in localizing the tumor. The classic neurilemmoma is a lobulated mass and orientated in an anteroposterior alignment within the orbit. The superior orbit is the most common site involved and the lesion is more frequently extraconal than intraconal.[4] However, some reported that the distribution of neurilemmomas in the retrobulbar space and the frequency in intra- and extrconal compartments appears random.[9] On MR imaging study, a more heterogenous appearance is noted in the neurilemmomas. On T2-weighted scans, these tumors are typically hyperintense to brain and isointense with cerebrospinal fluid. Marked enhancement with gadolinium is typical of neurilemmomas.[4]

Focal or generalized bony changes have been reported in four fifths of neurilemmomas.[21] Generalized enlargement of the orbit is more frequent with intraconal tumors. Frank erosion through the bone is seen only in tumors which were wholly or partially extraconal.[21] Meanwhile, bony expansion with enlargement of superior or inferior orbital fissures is not uncommon.[4] Widening of the superior orbital fissure is evident in almost a quarter of the neurilemmomas, with equal frequency for intra- or extraconal locations.[21] However, to our knowledge, the association of PSD with orbital neurilemmoma has not been reported.

PSD was first described by Benjamins[7] and was extensively reviewed by Lombardi *et al*.[8] It is characterized by progressive expansion of one or more paranasal sinuses associated with progressive thinning of the sinus wall, but without evidence of localized or mucous-membrane changes.[8] It may affect any of paranasal sinuses and most often affects the frontal sinus.[8] On CT scan, PSD may show an enlarged, aerated sinus with an abnormally thin wall, and is usually a benign asymptomatic process. However, it may cause progressive visual loss from optic nerve neuropathy if the sphenoid sinus involves.[16,18,22] PSD is diagnosed with standard radiography, CT scan or MR imaging study. The diagnostic criteria include (1) the enlargement of an air cell or the whole sinus, (2) the presence of only air in the abnormal space, and (3) the balloning outward of the walls of the sinus, which may be thinned and/or demineralized.[18]

The pathophysiology of PSD remains unclear. In normal subjects, there is a wide variation in the degree of pneumatisation of the sinuses. Overgrowth may occur in acromegalic subjects under the influence of a general growth factor or as a compensatory phenomenon in cases of agenesis of a cerebral hemisphere.[9,12,23] The underlying mechanism for a localized PSD is even more obscure with many speculative hypothesis. Several authors suggested a ball-valve like air-trapping mechanism in which failure of the air pressure within the sinus to equilibrate with the environment leads to sinus expansion.[18,24] However, in only one reported case has the stenosis of a sinus ostium been documented.[25] Congenital abnormality

has also been proposed as a possible mechanism that due to a congenital abnormality, unchecked development and growth of the sinus cavity and the meningeal cells leads to the PSD and associated tumors.[12,26] In PSD associated with pathologic processes such as meningioma and fibro-osseous diseases, some suggested that the meningioma or fibro-osseous diseases in some way stimulate the bone to bulge by mechanical or chemical factors.[12,17,23] Wiggli *et al*. suggested that chronic pressure might induce a bone remodeling process with predominant osteoblastic activity. Sinus expansion might occur at the bone remodeling place.[9] However, other authors suggested that the primary event is expansion of the sinus and that the expansion with its related thinning of bone stimulates the growth of adjacent meningeal cells to produce a meningioma.[8]

Sphenoid PSD was reported to be a sign of intracanalicular optic nerve sheath meningioma.[10-12] PSD associated with suprasellar meningioma,[9] with orbital fibro-osseous diseases[13-15] and with intracranial meningioma[17] has also been reported. This series found no case associated with neurofibromatosis, and no sign of fibro-osseous disorders in all patients. To our knowledge, the association of PSD with orbital neurilemmoma has not been reported yet. In our series, 4 cases (50%) of neurilemmoma were associated with PSD. The incidence of PSD in the orbital neurilemmomas was as high as 50% in our study. One patient was 60-year-old, with the rest being 24-32 years of age. The possibility of congenital abnormality as the underlying mechanism for the association of neurilemmomas with PSD, though unlikely, cannot be excluded. Three cases had the neurilemmomas in superior orbit, and one case in medial orbit. These tumors were solitary and well defined, which make the ball-valve like mechanism unlikely since they do not seem to occlude the ostium of the paranasal sinuses. All the neurilemmomas in these 4 cases were extraconal in location. Since the frank bony erosion occurs only in neurilemmomas with extraconal locations, we would expect a greater mechanical and/or chemical effect on the neighboring paranasal sinus from these extraconal neurilemmomas. The stimulating effect from the extraconal neurilemmonas possibly induces the paranasal sinus to develop over-pneumatization. Although the mechanism underlying the association between PSD and neurilemmoma is still unkonwn, from our observation, we suggest that pneumosinus dilatans may be a significant sign of orbital neurilemmoma.

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