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Pneumosinus dilatans after prolonged cerebrospinal fluid shunting in young adults with cerebral hemiatrophy. A report of two cases and review of the literature

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Abstract

Pneumosinus dilatans is a generalized or partial enlargement of the paranasal sinuses containing only air. Pneumosinus dilatans occures as an idiopathic disorder as well as in association with other disorders, including cerebral hemiatrophy. We report two cases of patients with congenital cerebral hemiatrophy who developed juvenile pneumosinus dilatans of the frontal, ethmoidal, and sphenoidal sinus. The hydrocephalus of both patients was treated by prolonged cerebrospinal fluid shunting. The development of hyperpneumatization of the paranasal sinuses was proved by plain radiographs and CT. Previous reports of pneumosinus dilatans are reviewed, and the effect of prolonged cerebrospinal fluid shunting in our cases is discussed. Cerebrospinal fluid shunting during the period of physiological pneumatization of paranasal sinuses might have increased hyperpneumatization.

Keywords: Cerebral hemiatrophy, cerebrospinal fluid shunting, frontal sinus, hydrocephalus, pneumosinus dilatans, sphenoid sinus.

1 Introduction

Pneumosinus dilatans is reported as a rare condition of pathological expansion of paranasal sinuses. Plain radiographs and CT show an enlargement of frontal, sphenoid, or ethmoidal sinuses containing only air. The alterations involved either all sinuses or only one. Commonly, the thin and demineralized wall of the involved sinus was ballooning outward, compressing neighbouring brain tissue. Upward bulging and hyperostosis of planum sphenoidale was called blistering [3, 24, 33, 34].

Prolonged cerebrospinal shunting procedures caused remodeling of the base and calvarium [17]. Two patients with cerebral hemiatrophy showed hyperpneumatisation of paranasal sinus after shunting for several years. Standard radiographs and CT reported the development of sinus expansion. The entity of pneumosinus dilatans is reviewed. Both cerebral hemiatrophy and ventricular shunting might have been responsible for enlargement of paranasal sinuses.

1 Case reports

Case 1

A 17-year-old boy developed internal hydrocephalus after having had a postnatal E. coli meningitis with complete blockage of aquaeductus Sylvii. At the age of four months a ventriculoatrial shunt was inserted. This shunt has been revised three times because of malfunction and infection. At the age of nine the drainage was converted into a ventriculoperitoneal shunt.

Generalized seizures occured within the first months. Antiepileptic drugs have kept the seizures completely under control for the last several years. Since childhood the patient suffered from severe tetraspasticity, oligophrenia, and increasing visual loss. Levels of prolactin were slightly raised and of of cortisol mildly depressed.

The early beginning of pneumatisation was followed by a severe bilateral enlargement of frontal, ethmoidal, and sphenoidal sinus. This was demonstrated from the fifth to the eleventh year shown by plain radiographs (Figure 1), and from the eleventh to the sixteenth year by CT (Figure 2). The walls of paranasal sinuses were thinned and blurred in CT. Hyperpneumatisation involved or-

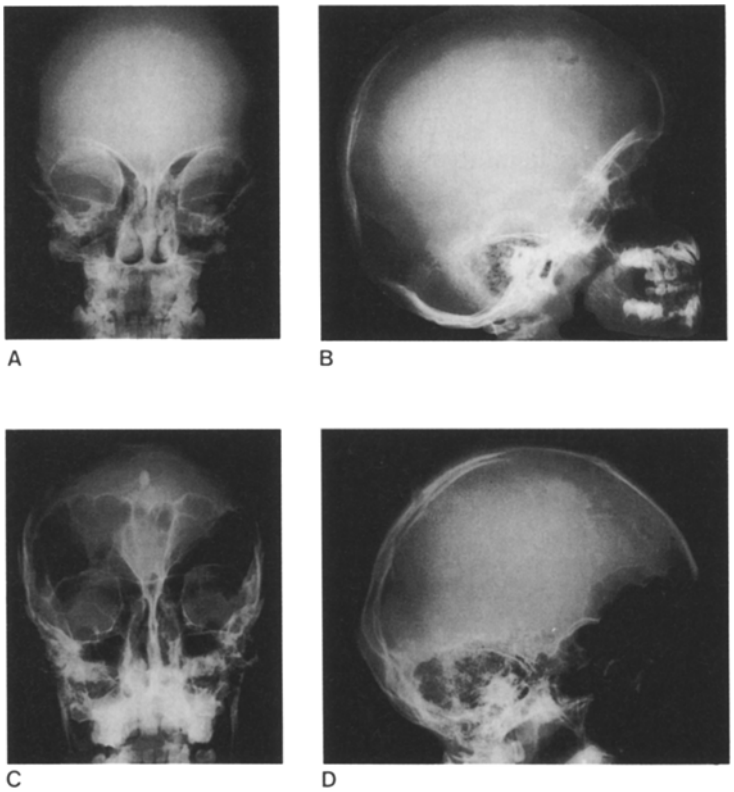

Figure 1. Case 1. **A, B:** at the age of 5 years: early beginning of a marked pneumatization of paranasal sinuses. **C, D:** at the age of 16 years: generalized pneumosinus dilatans of frontal, ethmoidal, and sphenoidal sinus, upward bowing and thickening of the planum sphenoidale, and reduction in the size of the sella turcica.

bital roof, clivus, clinoid processes and mastoid processes prefering cranial bones on the left side (Figure 2). The lateral view of the skull showed a remarkable thickening and upward bowing ("blistering") of the planum sphenoidale, and reduction of the size of sella turcica [37]. The brain structure was malformed with frontoparietal atrophia on the left slightly displacing midline structures to the right side and hypoplasia of the corpus callosum. CT revealed cerebellar dysplasia, particularly of the cranial part of vermis. The ventricular shunt was placed in the right lateral ventricle.

Case 2

A 16-year-old boy suffered from a congenital internal hydrocephalus of unknown genesis. Showing the clinical signs of raised intracranial pressure, he was provided with a ventriculoatrial shunt. Malfunctioning and shunt infection required multiple revisions. The atrial drainage was changed to peritoneal shunt at the age of five and the ventriculoatrial shunting system reinserted ten years later.

In the first year, EEG showed abnormality, but the patient did not show the clinical correlate of epilepsy. The first neurological examination proved severe tetraspasticity and oligophrenia, with preserved ability of speaking. Ophthalmoscopy showed no involvement of the optic disc; visus was preserved on both sides. The patient also suffered from a megacolon congenitum (M. Hirschsprung).

Radiological examination revealed progressive hyperpneumatisation of frontal, ethmoidal, and sphenoidal sinus beginning at the age of 11 and lasting until CT control at the age of 15 (Figures 3 and 4). In contrast to the first case, pneumosinus dilatans developed more asymmetrically, prefering the side of brain hemiatrophy. Hyperpneumatis-

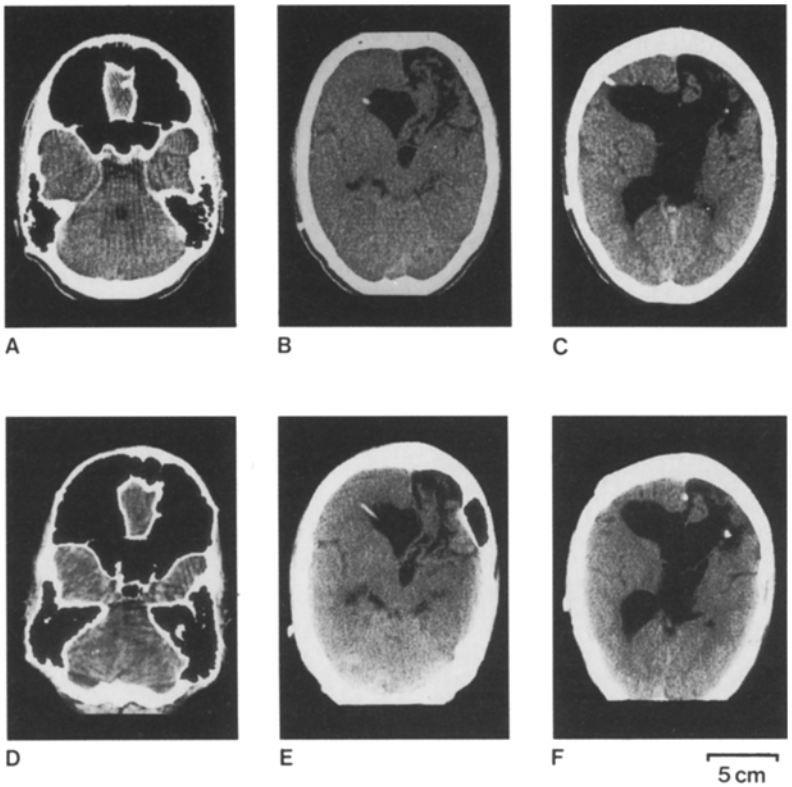

Figure 2. Case 1. **A–C:** CT at the age of 11 years: frontoparietal aplasia of the left hemisphere associated with dysplasia of the corpus callosum and cerebellum, generalized hyperpneumatization of paranasal sinuses. **D–F:** CT at the age of 15 years: increase of paranasal hyperpneumatization predominantly on the side of cerebral hemiatrophy.

ation extended to the orbital roof, clivus, and clinoid processes. The lateral view of the skull showed an upward convexity and hyperostosis ("blistering") of the planum spheniodale and a marked reduction in size of the sella turcica [37]. The cerebral hemiatrophy caused dilatation of the right lateral ventricle and ipsilateral displacement of the midline structures reducing the brain tissue to a 10 to 12 mm thin sheet. Associated malformations affected the corpus callosum with agenesis, the cerebellum with remarkable dysplasia, and upward shift of the tentorium. The ventricle shunt was positioned in the right lateral ventricle.

3 Discussion

A hyperpneumatisation of the paranasal sinuses, i.e., pneumosinus dilatans, has occured in association with heterogeneous diseases: cerebral hemiatrophy [4, 9, 14, 24, 33, 48], infantile meningoencephalitis [33, 36], psychomotor retardation [2, 3], acromegalia [2, 3, 46], neurofibromatosis generalisata (Recklinghausen's disease) [2, 3, 24], basal-cell naevus syndrome [2, 3, 24], dermatomal hemangiomas with cranial vascular malformation (Klippel-Trenaunay-Weber syndrome) [39], fibrous dysplasia [2, 3, 18, 24], cranial trauma [43], fractures of the planum sphenoidale [2, 3, 24], and meningiomas and other brain tumors [3, 5, 12, 13, 22, 24, 46].

In addition, 65 cases of idiopathic dilatation of paranasal sinuses have been reviewed in the literature. The frontal sinuses were involved in 39 cases, the sphenoidal sinuses in 15 cases, the maxillary sinuses in 12 cases, and the ethmoidal sinuses in 7 cases [1, 11, 20, 21, 22, 23, 25, 30, 32, 34, 38, 43, 45, 47, 49]. Pneumosinus dilatans frequently occured in male patients; the sex incidence showed a ratio of 53 male patients to 8 female patients. The age ranged from 13 to 76 years [20, 34].

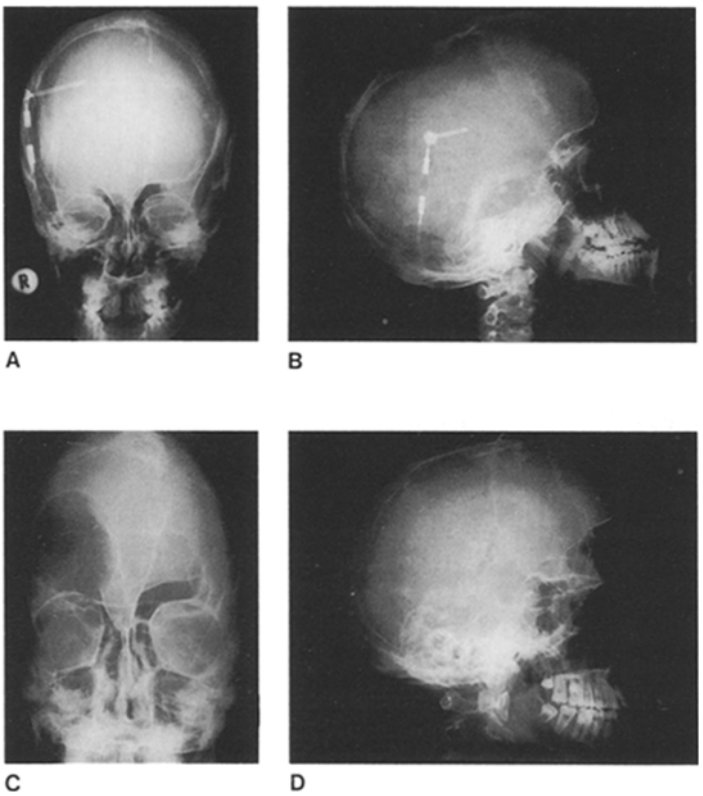

Figure 3. Case 2. **A, B:** at the age of 11 years: beginning of the pneumatization of the paranasal sinuses, ventricular shunt on the right side. **C, D:** at the age of 15 years: pneumosinus dilatans involving only paranasal sinuses on the side of cerebral hemiatrophy, upward convexity and hyperostosis and the planum sphenoidale, and small sella turcica.

The progressively expanding sphenoidal sinus behaved as does an osseous tumor, compressing the optic chiasm and narrowing the optic canal. The clinical symptoms included impaired visual acuity and vision loss on one or both sides, bitemporal hemianopsia, and optic atrophy. Impaired vision was observed in 12 out of 15 patients with dilating sphenoid sinus [20, 21, 34, 47]. Furthermore, headache and, in few cases, endocrine disorders like galactorrhea and mild hypopituitarism occured [3, 20, 34]. In our first case, we observed a progessive severe visual loss in both eyes, with an enormous enlargement of both sphenoidal issues (Figures 1 and 2). Compression of hypophyseal structures might have caused the mild prolactin increase and cortisol decrease [7]. Our second case proved, in agreement with Reicher et al. [34], that sphenoid sinus expansion did not necessarily impair vision.

In both patients CT accurately reflected the typical findings of cerebral hemiatrophy (Figures 2 and 4): atrophy and hypoplasia of one cerebral hemisphere, enlargement of the affected lateral ventricle, and ipsilateral displacement of the midline structures [4, 14, 48]. Furthermore, hypoplasia (Figure 2) and aplasia (Figure 4) of the corpus callosum were found in association with cerebellar anomalies resembling an Arnold-Chiari malformation (Figures 2 and 4). They showed reduction of posterior cranial fossa, cranial herniation of cerebellar structures through the tentorium, incomplete demarcation of the forth ventricle, and atypical midbrain configuration [26–28].

Agenesis of the corpus callosum was associated with an internal hydrocephalus in 40% of cases and with other malformations of the central nerv-

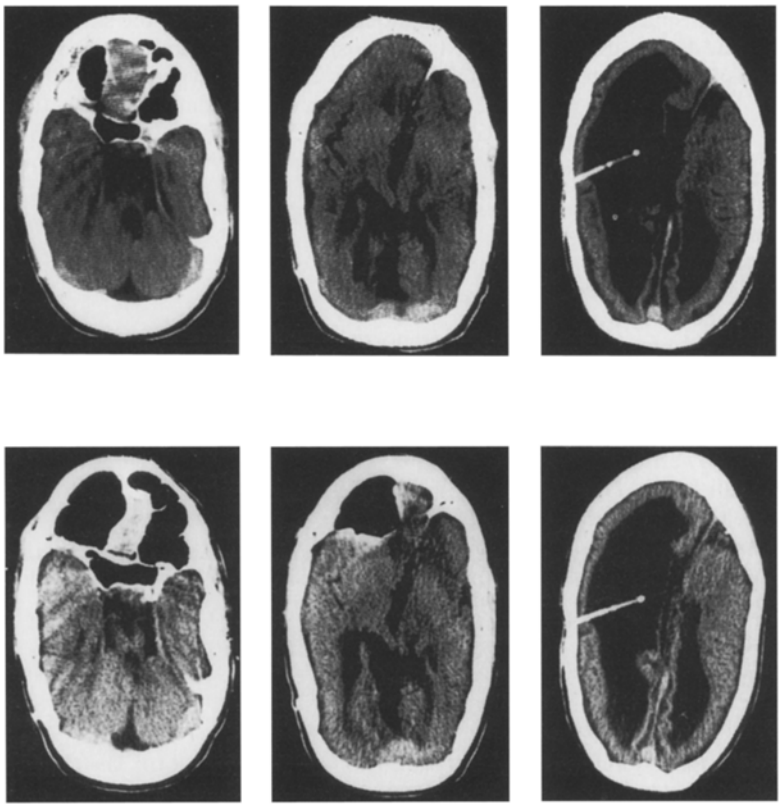

Figure 4. Case 2. **A—C:** CT at the age of 12 years: Cerebral hemiatrophy of the right hemisphere, aplasia of the corpus callosum, dysplasia of the cerebellum, hyperpneumatization of the right paranasal sinuses. **D—F:** CT at the age of 15 years: marked increase in the unilateral expansion of paranasal sinuses.

ous system up to 90% of cases [15, 16, 31]. Arnold-Chiari malformations, on the other hand, frequently caused internal hydrocephalus and occasionally combined with agenesis of the corpus callosum and other abnormalies of the brain [8, 10, 16]. Agenesis of the corpus callosum and associated malformations of the brain correlated with psychomotor retardation in 26—90%, with spastic paresis in 41%, and with epileptic seizures in 23—55% of the reviewed cases [16, 19, 44]. The clinical symptoms and radiological findings of both our patients reflected this pathological feature.

A hyperpneumatisation of the paranasal sinuses on the side of cerebral hemiatrophy has been previously described [4, 6, 14, 33, 35, 36, 48]. In our first case, radiographic imaging revealed a general dilatation which involved the contralateral sinuses (Figures 1 and 2). This might reflect the effect of various pathophysiological factors on the expansion of paranasal sinuses. Several children with brain hypoplasia and hydrocephalus required cerebrospinal fluid shunting. Commonly, the treatment caused no abnormal enlargement of paranasal sinuses. Thus the mechanism of developing pneumosinus dilatans has been, in some cases, undetermined.

In both cases the abnormal extension of paranasal sinuses occured during the physiologic period of rapid development in early puberty. The frontal and ethmoidal sinuses were commonly detectable on plain radiographs within the first two years, the sphenoid sinus at the age of three or four. The shape of the frontal sinus showed stronger phys-

iological variation than that of sphenoid sinus [24, 33]. Radiographic findings reflected the earlier and more prominent growth of paranasal sinuses in the first case (Figure 1).

An impaired development of the atrophic hemisphere decreasing the brain tissue pressure on surrounding cranial bone has been discussed as the cause of the expansion of paranasal sinus in cerebral hemiatrophy [29]. Decreased brain tissue pressure increases the local blood supply of the neighboring bone by facilitating the venous return. This, in turn enhances the growth and pneumatisation of the adjacent paranasal sinus [22, 29, 40, 41, 42]. It seemes likely that cerebrospinal fluid shunting could intensify this mechanism by decreasing the intracranial pressure and lowering the compression of the surrounding skull bone. The cerebrospinal fluid shunting might, therefore, be responsible for the generalized hyperpneumatisation of paranasal sinuses in the nonatrophic hemisphere in our first case. Kaufmann et al. [17] reported a remodeling of the base of the skull after prolonged cerebrospinal fluid shunting. They report an upward convexity and hyperostosis ("blistering") of the plenum sphenoidale and reduction in size of the sella turcica (Figures 1 and 3). We observed both of these abnormalities in both of our cases.

4 Conclusion

Previously reported cases as well as our own cases show that congenital cerebral hemiatrophy can initiate the development of juvenile pneumosinus dilatans. Cerebrospinal fluid shunting used to treat hydrocephalus caused by cerebral hemitrophy and associated malformations can aggreviate the hyperpneumatisation of paranasal sinuses.

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