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Anterior Transorbital Meningoencephaloceles: A Defect in the Pars Orbitalis of the Frontal Bone

Robin I. Davidson¹ and Paul K. Kleinman²

Anterior encephaloceles have been extensively reported and classified. Most have occurred at the juncture of the anterior part of the sphenoid with the pars orbitalis of the frontal bone. This corresponds to the region of either the crista galli or the cribriform plate. Encephaloceles in this location are seen in the region of the glabella, medial orbit, or in the nasal cavity or nasopharynx and have been described in detail [1]. Posterior orbital defects also occur and appear in conjunction with partial agenesis of the greater wing of the sphenoid bone, most frequently associated with neurofibromatosis [2, 3]. A more lateral anterior defect in the roof of the orbit involving the pars orbitalis of the frontal bone is extremely rare, and to our knowledge only one case has been briefly mentioned in the literature [4]. This form of anterior encephalocele has not been included in the comprehensive classification formulated by Suwanwela and Suwanwela [5].

We recently encountered two patients with congenital defects in the anterior part of the pars orbitalis of the frontal bone. This structure, derived from membranous bone, forms most of the roof of the orbit and the floor of the anterior fossa. On the basis of the embryogenesis of the orbital roof defects and the radiologic and gross anatomic features in these patients, we believe that this lesion should be considered an additional although rare form of anterior encephalocele. Thorough radiologic evaluation, including tomography, is necessary for correct diagnosis and for planning appropriate surgical management.

Case Reports

Case 1

A 4-year-old boy was referred after 1 year of progressive depression of the right eye associated with minimal proptosis. There was no diplopia or change in visual acuity. There had been minimal trauma to the right orbital region at 1½ years of age. There was no family history or neurocutaneous stigmata associated with neurofibromatosis.

A limited skull series 2 years before admission, after minimal frontal trauma, revealed elevation of the lesser wing of the right sphenoid bone. The current skull films demonstrated similar eleva-

tion of the right lesser wing of the sphenoid (fig. 1A). An optic foramen view (Reese projection) showed a 1.5 × 1 cm ovoid defect in the anterolateral roof of the right orbit (fig. 1B). The optic foramen and superior orbital fissure were intact. Oblique and lateral polytomography delineated the defect and revealed its thickened and sclerotic margins (figs. 1C and 1D). A coronal computed tomography (CT) scan demonstrated the orbital defect, as well as inferior and anterior displacement of the globe (fig. 1E). There were no associated intracranial abnormalities.

The patient had a right frontal craniotomy that entailed removal of the pars frontalis. Although the dura in the pars orbitalis defect extended into the orbit, a good cleavage plane could not be developed and the basal meningoencephalocele was transected. Excisional biopsy of residual intraorbital tissue revealed fibrillary gliosis. A synthetic dural substitute was secured to the basal dura and a tantalum/acrylic-mesh cranioplasty was attached to the defect in the pars orbitalis. His proptosis and ocular depression regressed and he continued asymptomatic.

Case 2

An 8-month-old boy was born with a right supraorbital defect, which was visible and palpable and associated with a proptosis and agenesis of the lateral two-thirds of his eyebrow. The scalp overlying this area was thickened with lipomatous tissue. There was no family history or neurocutaneous stigmata associated with neurofibromatosis.

An anteroposterior skull film revealed an obvious defect involving the right pars orbitalis and pars frontalis (fig. 2A). The entire right orbit was displaced inferiorly compared with the left. CT demonstrated the defect and evidence of herniated frontal lobe within it (figs. 2B and 2C). The right cerebral hemisphere was small with evidence of dilatation of the ipsilateral ventricular system and subarachnoid space.

At surgery, the frontal bone with its attendant supraorbital defect was removed. With extradural dissection, a line of demarcation between the dura and periorbita was developed and the meningoencephalocele was reduced through the defect in the pars orbitalis. A thick Silastic sling was used to create a prosthesis to form a floor for the anterior fossa. The supraorbital defect was closed by using a tantalum mesh/methyl-methacrylate cranioplasty. The child's postoperative course was benign and he underwent one subsequent revision for cosmetic reduction of a lateral irregularity in the superior skull defect.

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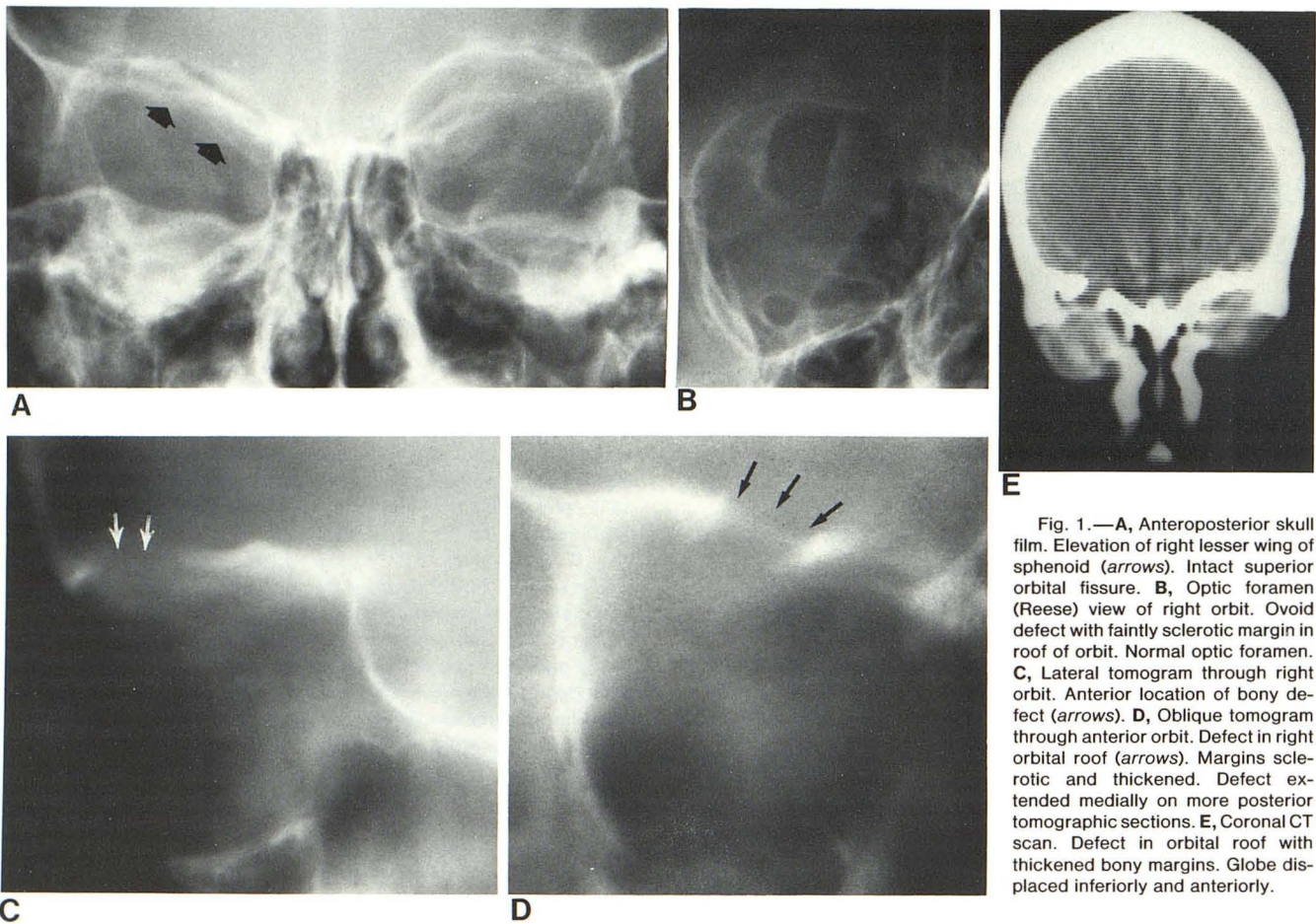


Fig. 1.—A, Anteroposterior skull film. Elevation of right lesser wing of sphenoid (arrows). Intact superior orbital fissure. B, Optic foramen (Reese) view of right orbit. Ovoid defect with faintly sclerotic margin in roof of orbit. Normal optic foramen. C, Lateral tomogram through right orbit. Anterior location of bony defect (arrows). D, Oblique tomogram through anterior orbit. Defect in right orbital roof (arrows). Margins sclerotic and thickened. Defect extended medially on more posterior tomographic sections. E, Coronal CT scan. Defect in orbital roof with thickened bony margins. Globe displaced inferiorly and anteriorly.

Discussion

The anterior sphenoidal anlage, forming the midline and paramedian structures of the floor of the anterior fossa, develop by endochondral ossification [6, 7]. The frontal bone, developing by intramembranous ossification, is identifiable as a bilobar primary membranous ossification center in the position of the future frontal eminence at gestation day 40–50. It appears initially as two finely reticulated areas of ossification which together constitute the single primary ossification center. The long axis of the center corresponds to the position of the superciliary arch, which then develops to form initially the lower part of the frontal squamae. The trabecular development of membranous ossification then progresses from the superciliary ridge into the pars orbitalis [6, 7].

The overall incidence of anterior encephaloceles in non-Asiatic countries is difficult to determine. The 1943 article of Ingraham and Swan [8] classified basal encephaloceles into sphenopharyngeal, sphenoorbital, sphenothmoidal and transtethmoidal, and sphenomaxillary. Of 546 patients with spina bifida and cranium bifidum, they encountered eight patients with basal encephaloceles, five of which were transsphenoidal and three transtethmoidal. The 1970 incidence figures of Nealy et al. [9], involving a 20 year experience with children with encephaloceles and myelomeningo-

coceles, list 559 patients with myelodysplasia, 60 with encephaloceles, and four with both lesions. Of these 64 patients, 54 lesions were occipital, three were interparietal, eight were nasofrontal, and one was nasopharyngeal. Of the nasofrontal lesions, they state that in one case "the right frontal region and orbit were involved."

Extensive literature exists dealing with anteromedial meningoencephaloceles ("sincipital encephaloceles") [1, 5, 10–17]. This literature does not reveal any description of a defect analogous to our two patients, with the exception of the single case of Dandy [4] in his treatise *The Brain*. This male infant with a large orbitofrontal encephalocele is similar to our case 2 and was treated with a split parietal bone transplant. The description of Dandy of the procedure to repair orbital defects, illustrated for a patient with a posterior orbitosphenoid defect, is reminiscent of the approach used in our two patients [18].

A classification of sincipital meningoencephaloceles was provided in 1972 by Suwanwela and Suwanwela [5] reporting from Thailand where anterior defects occur with greater frequency than in the Western Hemisphere. Their categorization includes the lesions noted in table 1, and does not include a description of a defect in the pars orbitalis.

Other articles describe the defects that occur commonly with systemic neurofibromatosis [2, 3, 5]. In this condition the posterior half of the roof of the orbit is absent in asso-

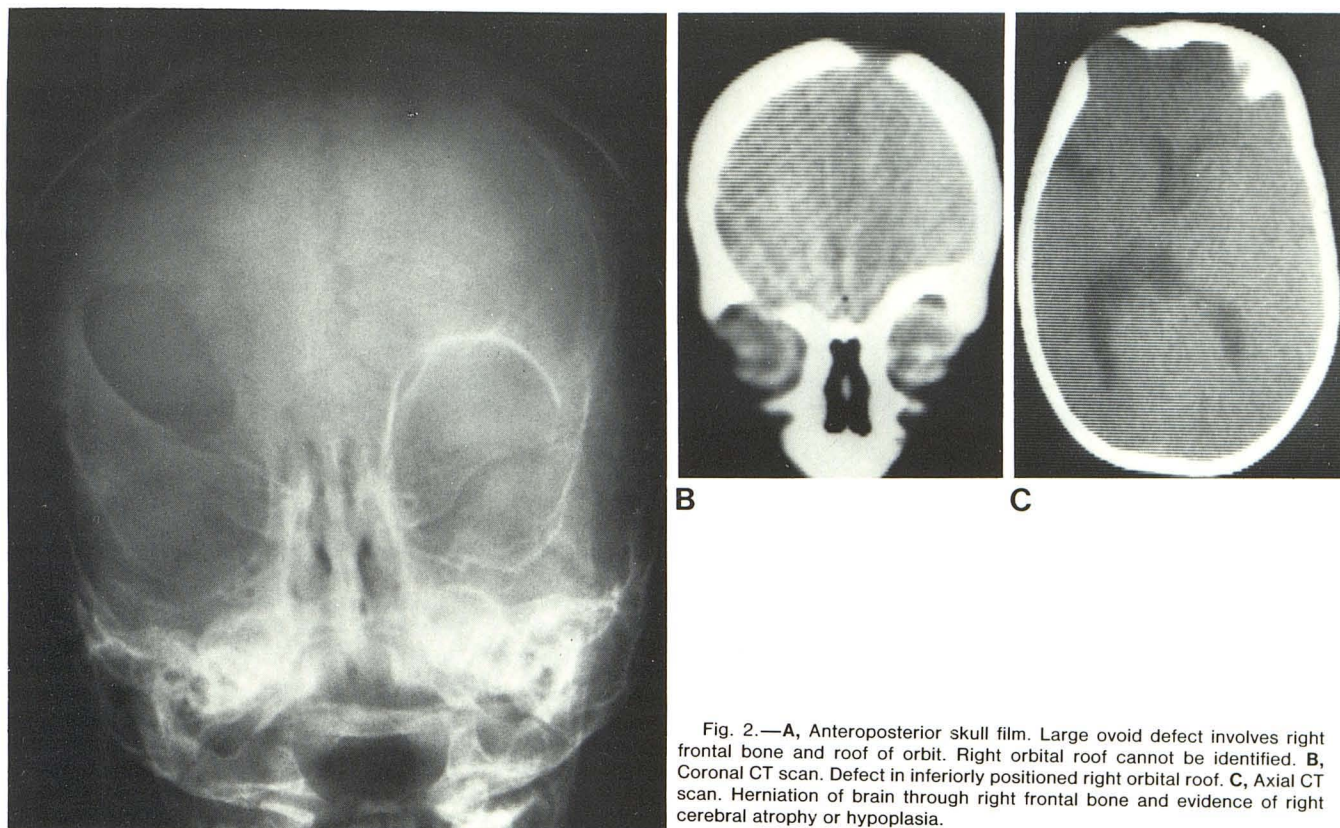


Fig. 2.—A, Anteroposterior skull film. Large ovoid defect involves right frontal bone and roof of orbit. Right orbital roof cannot be identified. B, Coronal CT scan. Defect in inferiorly positioned right orbital roof. C, Axial CT scan. Herniation of brain through right frontal bone and evidence of right cerebral atrophy or hypoplasia.

A

TABLE 1: Classification of Encephalomeningoceles*

Type
Occipital
Of the cranial vault:
Interfrontal
Anterior fontanel
Interparietal
Posterior fontanel
Temporal
Frontoethmoidal:
Nasofrontal
Nasoethmoidal
Nasoorbital
Basal:
Transethmoidal
Sphenoethmoidal
Transsphenoidal
Frontosphenoidal or sphenoorbital
Transorbital
Cranioschisis:
Cranial (upper facial cleft)
Basal (lower facial cleft)
Occipitocervical cleft
Acrania and anencephaly

* Adapted from [5].

ciation with the greater wing of the sphenoid. Hence the posterior orbit is pushed anteriorly by posterobasal dura of the frontal lobe and by anterior middle fossa contents. This lesion was well illustrated by Lewald [3] and Fargueta et al. [2]. The latter authors also described the nasofrontal orbital

encephalocele but not a more lateral defect in the pars orbitalis.

The probable site of the defect responsible for the bulk of sincipital meningoencephaloceles is at the point of juncture of the anterior sphenoidal chondrocranium with the developing membranous bone of the pars orbitalis of the frontal bone. In our patients, it appears that a part of the primary membranous ossification center for the frontal bone failed to develop a component of the superciliary ridge and pars orbitalis. The etiology of this developmental arrest in membranous ossification, occurring at about 42 mm crown rump length, 55–60 days gestation, is obscure. In our patients it resulted in a nonpulsatile proptosis and depression of the orbital contents in both instances and an accompanying visible supraorbital defect in one patient (fig. 3).

Several entities should be considered in the differential diagnosis. The more common variety of orbital encephalocele has a medial or posterior direction. The detailed radiographic description of paramedian anterior meningoencephaloceles of Harverson et al. [19] defined the defect as occurring between the frontal and ethmoidal bone at the site of the foramen cecum. This clearly contrasts with the anterolateral defects in our cases.

Patients with neurofibromatosis commonly demonstrate a dysplastic sphenoid bone complex that may include elevation of the lesser wing of the sphenoid, a finding present in case 1. However, enlargement of the superior orbital fissure, hypoplasia of the sphenoid wings, and downward tilting of the floor of the sella, which occur in neurofibromatosis, were

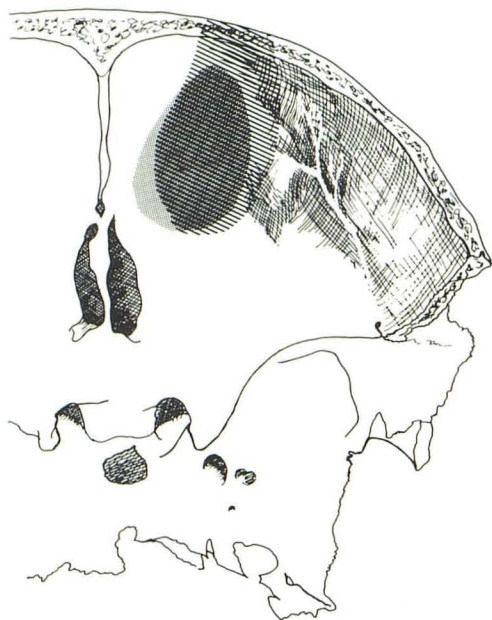


Fig. 3.—Floor of anterior fossa viewed from above. Schematic representations of both defects superimposed: Case 1, stippled; case 2, cross-hatched.

not present in these cases. When cerebral tissue protrudes into the orbit through a posterior orbital defect (commonly seen in association with neurofibromatosis), anterior middle fossa contents can be present intraorbitally [2, 18]. As the defects were anterosuperior in our patients, the encephalocoeles involved the orbital gyri of the basal frontal lobe.

An orbital roof fracture with leptomenigeal cyst formation enters the differential diagnosis, but the bony defect in such cases would be expected to be more linear with irregular expansion, rather than the smooth ovoid configuration seen in our patients. Computed tomography might also show evidence of associated intracranial pathology (e.g., a chronic subdural hematoma). A posttraumatic carotid cavernous fistula may be associated with pulsatile exophthalmos and a posterior orbital defect. However, this defect is related to enlargement of the superior orbital fissure due to a dilated superior orbital vein. Lastly, an intracranial neoplasm can erode through the orbital wall, and appear within the orbit with proptosis [20]. CT would exclude this possibility.

Thorough radiologic evaluation is essential in the diagnosis and treatment of all orbital encephalocoeles [3, 14, 19, 21]. Plain films and conventional tomography will delineate the size, configuration, and location of the bony defect. CT scanning will show any associated intracranial abnormality as well as the extent of cerebral herniation. CT after the subarachnoid administration of metrizamide may delineate a subarachnoid communication with the encephalocoele. Arteriography and pneumoencephalography have been performed in the evaluation of encephalocoeles, but with the availability of CT, they are probably unnecessary in most cases.

Treatment is effected by surgery in order to prevent further proptosis, diplopia, and possible amblyopia ex-

opsia. Removal of the right frontal bone and extra- or intradural reduction or resection of the meningoencephalocoele can generally be carried out without difficulty.

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