

Case Report

Posttraumatic Giant Extradural Intradiploic Epidermoid Cysts of Posterior Cranial Fossa : Case Report and Review of the Literature

Yavor Enchev, M.D., Ph.D., Bogidar Kamenov, M.D., Alla William, M.D., Vasil Karakostov, M.D., Ph.D.

Department of Neurosurgery, Medical University-Sofia, Sofia, Bulgaria

We reported a unique case of posttraumatic giant infratentorial extradural intradiploic epidermoid cyst. A 54-year-old male, with a previous history of an open scalp injury and underlying linear skull fracture in the left occipital region in childhood, presented with a painful subcutaneous swelling, which had been developed gradually in the same region and moderate headache, nausea, vomiting and cerebellar ataxia. The duration of symptoms on admission was 3 months. Imaging studies revealed occipital bone destruction and giant extradural intradiploic lesion. The preoperative diagnosis was giant infratentorial extradural intradiploic epidermoid cyst. Surgery achieved total removal of the lesion, which was histologically confirmed and the postoperative course was uneventful. To our knowledge, this is the first case of giant infratentorial extradural intradiploic epidermoid cyst with a traumatic etiology described in the literature.

Key Words : Neoplasm · Epidermoid cyst · Cranial fossa · Posterior · Trauma · Etiology

INTRODUCTION

Epidermoid cysts are benign, slow growing lesions, representing about 1% of all intracranial tumors^{1,2,5-7,9,10}. Intracranial epidermoids are subdivided to more frequent intradural and less common extradural subgroups¹⁰. Extradural epidermoid cysts are intradiploic in approximately 25% of the cases, and predominantly supratentorial in location^{1,5,7,10}. Infratentorial intradiploic epidermoids are not rare whereas the giant variants are extremely rare. We report an unique case of posttraumatic giant infratentorial extradural intradiploic epidermoid cyst. To the best of our knowledge, this is the first case of giant infratentorial extradural intradiploic epidermoid cyst with a traumatic aetiology described in the literature. Review of the literature is presented relevant to this unusual case along with their epidemiology, clinic, diagnosis, surgical treatment and etiology.

CASE REPORT

A 54-year-old man experienced bicycle accident with an open

scalp injury and underlying linear skull fracture in the left occipital region at 5 years of age. The wound had been treated surgically with lavage and sutures. Subsequently, painless subcutaneous swelling gradually developed in the same region. The patient has been well and without any complaints until 3 months before his admission in our clinic. Local inspection exposed a painful subcutaneous swelling, 7 to 12 cm across, in the occipital region, predominantly on the left side. Physical examination revealed headache, nausea, vomiting and cerebellar ataxia. Computed tomography (CT) exposed a giant infratentorial extracerebellar hypodense lesion with extensive occipital bone destruction and substantial mass effect in the posterior cranial fossa (Fig. 1A). Magnetic resonance imaging revealed the giant infratentorial extradural intradiploic tumor, inhomogeneously hypointense in T1-weighted images and hyperintense in T2-weighted, with an enhancement rim of the thickened dura mater and significant compression of the cerebellum, brain stem, fourth ventricle and the left occipital lobe, without corresponding brain edema (Fig. 1B). The preoperative diagnosis was giant infratentorial extradural intradiploic epidermoid cyst.

Under a general anesthesia, mass removal was performed starting with "Hockey-stick" skin incision. The exposed occipital bone was widely destroyed, thinner and partially perforated by the tumor (Fig. 1C), which was soft, whitish and cheesy. The dura mater was thickened but intact throughout (Fig. 1D). The tumor was totally removed including its capsule (Fig. 1E). The histological

• Received : April 19, 2010 • Revised : December 1, 2010
• Accepted : December 31, 2010
• Address for reprints : Yavor Enchev, M.D., Ph.D.
Department of Neurosurgery, Medical University-Sofia, 15, Boulv. "Acad. Ivan Geshov", 1431 Sofia, Bulgaria
Tel : +359-888 441191, Fax : +359-9294-5492
E-mail : dr.y.enchev@gmail.com

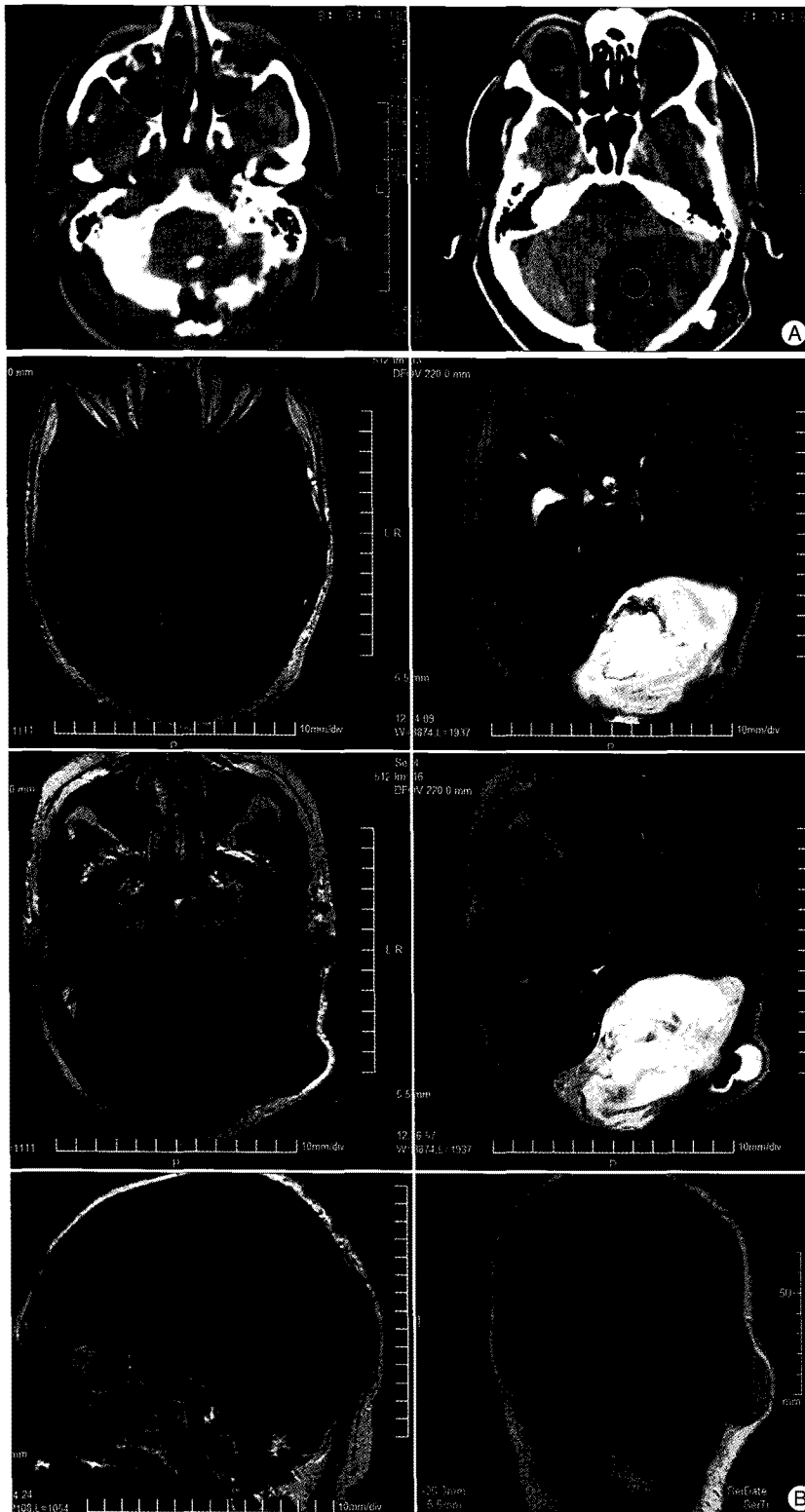


Fig. 1. Images of posttraumatic giant intradiploic epidermoid cyst of posterior cranial fossa in the present case. A : Preoperative computed tomographic (CT) scans show giant infratentorial extracerebellar hypodense lesion with extensive occipital bone destruction and substantial mass effect in the posterior cranial fossa. B : Preoperative magnetic resonance (MR) images show giant infratentorial extradural intradiploic tumour, inhomogeneously hypointense in T1-weighted images (left) and hyperintense in T2-weighted (right), with an enhancement rim of the thickened dura mater and significant compression of the cerebellum, brain stem, fourth ventricle and the left occipital lobe, without corresponding brain oedema.

examination confirmed an epidermoid cyst and the postoperative course was uneventful.

DISCUSSION

Epidermoid cysts have expansive type of growth and cranial or spinal localization. Cranial epidermoid cysts are relatively rare (0.3-1.8% of all surgically treated craniocerebral tumors), benign and predominantly-intradural lesions¹⁰. Extradural epidermoid cysts represent about 25% of all cranial epidermoids and engage the scalp or the skull⁷. Extradural epidermoids of the skull are also known as intradiploic. The first primary intradiploic epidermoid cyst was reported by Müller in 1838⁸. Ciappetta et al.³ cited a total of 223 cases of intradiploic extradural epidermoids, reported in the literature by 1990. Some of these epidermoid cysts may attain giant size before they are diagnosed. The giant intradiploic extradural epidermoid cysts are rather uncommon (about 30 described cases)⁷ with a supratentorial predilection for the frontal and parietal bones of the skull^{6,11}.

The first case of infratentorial giant intradiploic extradural epidermoid cyst was reported by Rengachary et al. in 1978. To the best of our knowledge, there have been only 8 cases of such epidermoid in the literature, including the presenting case (Table 1).

The rate of epidermoids growing is slow, linear in contrary to the most of the other tumors with their exponential growth¹. In correspondence with that the age at the onset of complaints in this group (mean age of 55 years, range- 24-74 years) was logically higher compared with that of the non-giant intradiploic epidermoid cysts of the skull (mean age of 32 to 38 years)³. An utter male sex predilection was outlined in the current series (male : female ratio-8 : 0) in contrast to the data of other reviews of intradiploic epidermoid cysts of the skull^{3,6}.

The duration of symptoms on admission in the studied group was short and range between 1 and 5 months (mean 3 months), which is most likely explained

by the giant size of the lesions and their significant compressive effect. The presence of painful or painless subcutaneous swelling was not a compulsory, but extremely indicative feature of the diagnosis infratentorial giant intradiploic extradural epidermoid cyst^{3,6}. Neurological examination was non-uniformly positive depending on the predominant direction of the tumor growth- intra- or extracranial and the grade of the occipital bone destruction. In both cases with neurologically intact patients, the epidermoid cysts widened the space of the posterior cranial fossa^{5,7}.

The imaging diagnosis of the infratentorial giant intradiploic extradural epidermoid cysts does not represent a challenge. X-ray films of the skull and their radiation burden must be avoided, because the lytic occipital bone defect revealed by them is clearly visualized by CT, which in addition demonstrated the giant hypodense lesion and sometimes post-contrast rim enhancement of the thickened dura mater. Regardless that the surgery could be performed on only based on CT scans⁶, if available MR imaging should be obligatory in the preoperative investigations, because of its detailed imaging information^{1,7}. MR imaging reveal giant extradural lesion inhomogeneously hypointense in T1-weighted and hyperintense in T2-weighted images, with a post-gadolinium dural enhancement over the cerebellum. Magnetic resonance angiography was never performed in the series, but could substitute the conventional angiography with its hazards for the patients, in order to evaluate the grade of compression of the neighbouring dural venous sinuses.

The differential diagnosis of the infratentorial giant intradiploic extradural epidermoid cysts is quite limited, including dermoid cysts, eosinophilic granulomas, hemangiomas and in some cases-large arachnoid cysts⁷ and is easy to be solved.

Despite the huge size of the infratentorial giant intradiploic extradural epidermoid cysts the golden standard of the surgery is the total removal of the tumor with its capsule and preserving the integrity of the dura and its venous sinuses^{1,2,5-7,9,10}. Neuro-navigation could be useful in the cases with a predominantly intracranial growing and supposed invasion of the dural venous sinuses in order to limit the extent of craniectomy and to pre-

serve the sinuses integrity⁴. The total removal of the giant epidermoid cysts leads to permanent cure. Cranioplasty may be needed, when there is a large bony defect. In the series, the etiology of epidermoids was congenital sequestration of ectodermal cells within the cranial bones between the third and fifth embryonic week^{1,2,5-7,9,10,12}, except in the presented case where the inclusion of ectodermal cells thought to be occurred at the time of trauma.

CONCLUSION

We report an unique case of posttraumatic giant infratentori-

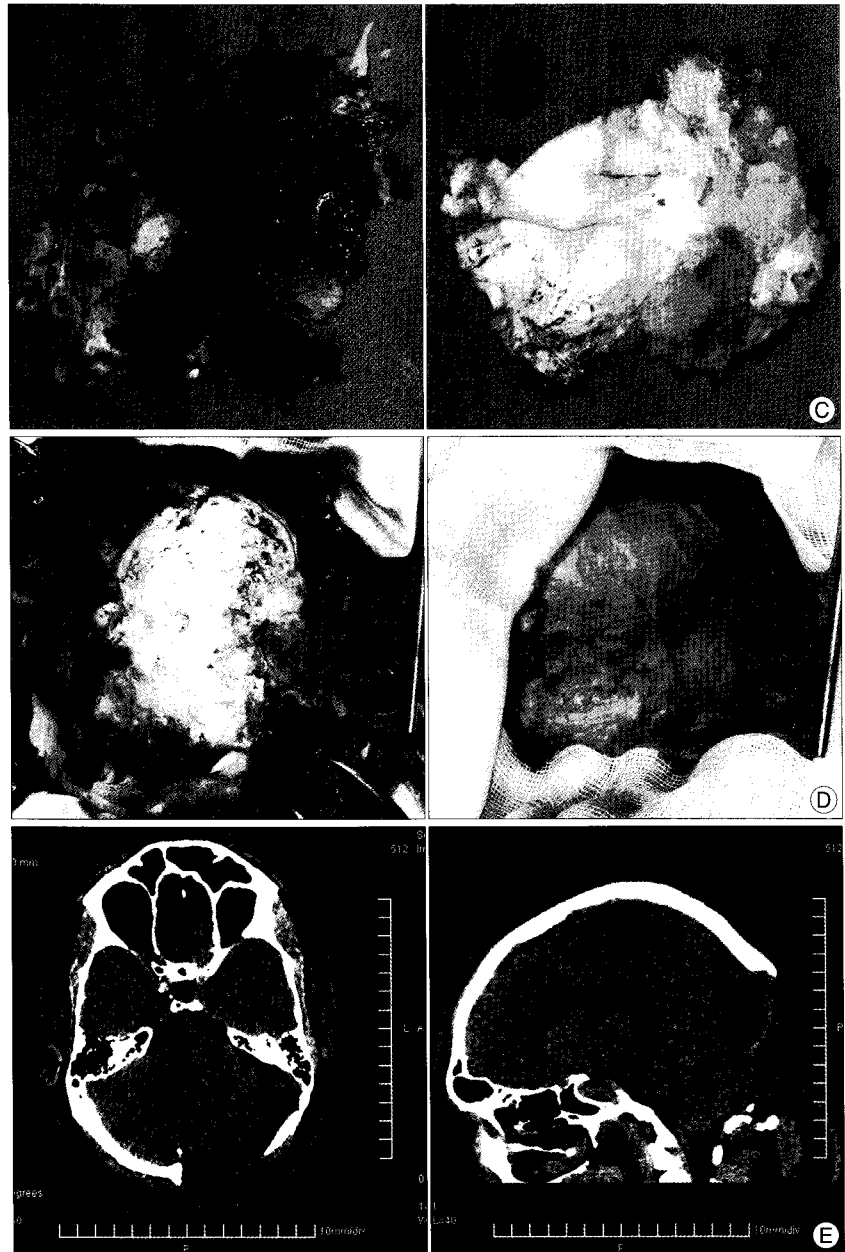


Fig. 1. Continued. C : Note the removed occipital bone which is widely destroyed, thinner and partially perforated by the tumour. D : Note the tumor which is soft, whitish and cheesy (left). The dura mater over the cerebellum is thickened and intact throughout (right). E : Postoperative CT scans one day after the procedure show total removal of the tumor.

Table 1. Cases with giant extraaxial intradiploic epidermoid cysts of posterior cranial fossa reported in the literatures up to date

No.	Authors (year)	Age (yrs)	Sex	Duration of symptoms on admission (mths)	Local state	Neurological state	X-ray	CT	Imaging diagnosis			Sur-gary	Supposed aetiology	
									Conventional angiography	T1-weighted	T2-weighted +gadolinium			
1	Rengachary et al. ⁹⁾	62	M	NM	Negative	Headache, nausea, vomiting, intermittent choking spells and nocturnal stridor	Lytic lesion of the occipital bone with expansion and thinning of the inner and outer tables	Hypodense	Obliteration of the distal superior sagittal sinus, transverse sinuses and torcular collateral venous drainage to veins of Labbe	ND	ND	ND	TR	Congenital
2	Rubin et al. ¹⁰⁾	27	M	2	Negative	Headache, occasional diplopia, papilloedema	Bone erosion	Isodense, calcification, rim enhancement	Compression of the distal superior sagittal sinus, right transverse sinus and torcular	ND	ND	ND	TR	Congenital
3	Guridi et al. ⁵⁾	47	M	1	Subcutaneous swelling	Negative	Homogeneous radiolucency, lytic occipital bone defect, sharply defined sclerotic borders	Hypodense	Compression without obstruction of the distal superior sagittal sinus, right transverse sinus and torcular	ND	ND	ND	TR	Congenital
4	Jaiswal et al. ⁶⁾	40	M	NM	Subcutaneous swelling	Cerebellar ataxia	Lytic occipital bone defect	Hypodense, occipital bone destruction		ND	ND	ND	TR	Congenital
5	Maiuri et al. ⁷⁾	60	M	5	Subcutaneous bone swelling, local pain	Negative	ND	Hypodense, occipital bone destruction		ND	Hyperintense inhomogeneously	Hyperintense inhomogeneously	TR	Congenital
6	Borha et al. ²⁾	73	M	4	Subcutaneous bone swelling, local pain	Headache, cerebellar ataxia, dysmetria	ND	Hypodense		ND	ND	ND	TR	Congenital
7	Alberione et al. ¹⁾	74	M	NM	Negative	Cerebellar ataxia, dysmetria	ND	Hypodense, calcification, rim enhancement		ND	Hyperintense inhomogeneously	Hyperintense inhomogeneously	TR	Congenital
8	Enchev et al. (the presented case)	54	M	3	Subcutaneous swelling, local pain	Headache, nausea, vomiting, cerebellar ataxia	ND	Hypodense, occipital bone destruction		ND	Hyperintense inhomogeneously	Hyperintense inhomogeneously	TR	Trauma

CT : computed tomography, NM : not mentioned by the authors, m : male, MRI : magnetic resonance imaging, mths : months, ND : not done, TR : total removal, yrs : years

al extradural intradiploic epidermoid cyst with an established traumatic etiology. The infratentorial giant extradural intradiploic epidermoid cysts are exceptionally rare, extremely slow growing, benign lesions, exclusively in males, with typical X-ray, CT and MRI findings and characteristic dormant clinical course. The non-complicated total removal of these lesions is associated with a good long-term prognosis with permanent cure and lack of recurrence at the follow-up. Caution should be paid in male patients, with an open scalp injury and underlying linear skull fracture in the occipital region and MRI-based follow-up is recommended annually for several years after the trauma, aiming in earlier diagnosis and timely surgical treatment of eventual posttraumatic infratentorial extradural intradiploic epidermoid cysts.

References

1. Alberione F, Caire F, Fischer-Lokou D, Gueye M, Moreau JJ : [Giant intradiploic infratentorial epidermoid cyst]. *Neurocirugia (Astur)* 18 : 423-426, 2007
2. Borha A, Emery E, Khouri S, Hitier M, Derlon JM : [Giant extradural epidermoid cyst of the posterior fossa]. *Neurochirurgie* 51 : 599-603, 2005
3. Ciappetta P, Artico M, Salvati M, Raco A, Gagliardi FM : Intradiploic epidermoid cysts of the skull : report of 10 cases and review of the literature. *Acta Neurochir (Wien)* 102 : 33-37, 1990
4. Enchev Y : Neuronavigation : geneology, reality and prospects. *Neurosurg Focus* 27 : E11, 2009
5. Guridi J, Ollier J, Aguilera F : Giant intradiploic epidermoid tumor of the occipital bone : case report. *Neurosurgery* 27 : 978-980, 1990
6. Jaiswal AK, Mahapatra AK : Giant intradiploic epidermoid cysts of the skull. A report of eight cases. *Br J Neurosurg* 14 : 225-228, 2000
7. Maiuri E, Del Basso De Caro M, D'Acunzi G, Tortora F, Esposito F : Giant intradiploic epidermoid cyst of the occipital bone. *Zentralbl Neurochir* 65 : 32-35, 2004
8. Müller J : Über den feineren Bau und die Formen der krankhaften Geschwülste, ed 1. Berlin : G. Reimer, 1838, pp73-81, German
9. Rengachary S, Kishore PR, Watanabe I : Intradiploic epidermoid cyst of the occipital bone with torcular obstruction : case report. *J Neurosurg* 48 : 475-478, 1978
10. Rubin G, Scienza R, Pasqualin A, Rosta L, Da Pian R : Craniocerebral epidermoids and dermoids. A review of 44 cases. *Acta Neurochir (Wien)* 97 : 1-16, 1989
11. SKANDALAKIS JE, GODWIN JT, MABON RF : Epidermoid cyst of the skull; report of four cases and review of the literature. *Surgery* 43 : 990-1001, 1958
12. Toglia JU, Netsky MG, Alexander E Jr : Epithelial (epidermoid) tumors of the cranium. Their common nature and pathogenesis. *J Neurosurg* 23 : 384-393, 1965