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A Case Report: A Non-infantile Desmoplastic Astrocytoma

Olgu Sunumu: İnfantil Olmayan Dezmoplastik Astrositom

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ABSTRACT

INTRODUCTION and CASE DESCRIPTION: Desmoplastic infantile astrocytomas (DIA) are rare intracranial tumors of infancy with distinctive clinical and radiological features. Despite their radiologically aggressive appearance they tend to follow a benign course with favorable prognosis even after subtotal resections. Non-infantile cases are rarely encountered with only four cases reported before. The authors present a non-infantile DIA in a 4 years old female patient and discuss the clinical features, diagnosis, treatment of this rare entity.

CONCLUSION: Although accepted as a tumor of infancy, DIA can also be encountered in older patients. Careful diagnosis and differentiation of DIA cases with other tumors, particularly malignant astrocytomas is important since the therapeutic strategies may differ.

KEY WORDS: Brain tumor, Desmoplastic infantile astrocytoma, MRI, Noninfantile, Pathology, Surgical treatment

ÖZ

Giriş ve olgu tanımı: Dezmoplastik infantil astrositomlar (DİA) infantil dönemde görülen ve farklı klinik ve radyolojik özellikleri olan intrakranial tümörlerdir. Radyolojik olarak çok agresif görünümlü olsalar da subtotal eksizyonlardan sonra bile iyi prognozlu, selim bir gidişat göstermektedirler. İnfantil olmayan olgulara çok nadiren rastlanır ve literatürde 4 olgu bildirilmiştir. Yazarlar bu yazıda 4 yaşındaki bir kız hastada infantil olmayan bir DIA sunmakta ve bu nadir olgunun klinik özelliklerini, teşhis ve tedavisini tartışmaktadır.

SONUÇ: İnfantil dönemin tümörü olarak kabul edilse de DIA olgularına daha ileri yaşlarda da rastlanabilir. Tedavi stratejileri farklı oldugu için DIA olgularının dikkatli teşhisi ve özellikle habis astrositomlardan olmak üzere diğer tümörlerden ayrımı çok önemlidir.

ANAHTAR SÖZCÜKLER: Beyin tümörü, Dezmoplastik infantil astrositom, MR, İnfantil olmayan, Patoloji, Cerrahi tedavi

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INTRODUCTION

Desmoplastic infantile astrocytomas (DIA) are rare supratentorial tumors of infancy, defined by Taratuto et al. in 1984 (10). Almost all patients present in the first 24 months of life having supratentorial large masses with cystic and solid components (11). Only four cases of DIA have been reported in non-infantile patients (1,2,4,5). We report an additional non-infantile DIA in a 4-year-old female and review the clinical features, histopathology, diagnostic methods and treatment of this rare entity.

CASE REPORT

A 4-year-old girl presented with a 2 days history of headache, vomiting, dizziness and tiredness. Questioning of the parents elicited that she was not willing to play with her friends, had attention deficit, and refused to use her left hand, all of which started 1 week prior to admission. On neurological examination, she was drowsy with no focal neurological signs or papilledema except for mild left global hemiparesis. Cranial magnetic resonance imaging (MRI) with and without gadolinium revealed a right parietooccipital heterogenous enhancing cystic mass (dimensions, 8x7x6,5 cm) with a midline shift of ~ 1,5 cm (Figure 1A,B,C). Following a right reverse question mark incision starting from tragus and extending posteriorly above the level of pinna, a temporoparietooccipital craniotomy is done. The tumor was firm, harboring cystic components and partially invaded the dura. The tumoral mass was grossly totally removed using the ultrasonic aspirator (CUSA).

Histopathological Examination

The dura-related cystic tumor showed a biphasic pattern consisting of spindle or round shaped tumoral cells with some plump nuclei which were glial fibrillary acidic protein (GFAP)-positive astrocytes, embedded in a desmoplastic stroma (Figure 2A). The tumor presented frequently wavy or storiform pattern, that was mimicking mesenchymal tumor (Figure 2B). The astrocytic cells had some pleomorphism, although the mitotic activity was very low. Scattered islands of undifferentiated small cells were seen acquiring GFAP positivity at their peripheral zone facing the collagen. (Figure 2C). It invaded surrounding brain tissue, usually along the blood vessels. The few small necrotic focus were observed. The tumor cells also were diffuse positive for vimentin and mostly S-100. No neuronal cell was observed on H+E sections or immunohistochemically. MIB-1 index of the tumor was established very low. These morphological findings were compatible with a desmoplastic infantile cerebral astrocytoma.

The postoperative period was uneventful and the patient was discharged at the 5th postoperative day without additional neurological deficit. Postoperative follow-up cranial MRI demonstrated near total removal of the tumor (Figure 3A,B,C). At the 11th follow-up, the patient was free of any signs or symptoms.

DISCUSSION

DIA's are extremely rare intracranial neoplasms of infancy and only 4 noninfantile cases have been reported in the literature (1,2,4,5). The demographic features of the previously reported cases are summarized in (Table I).

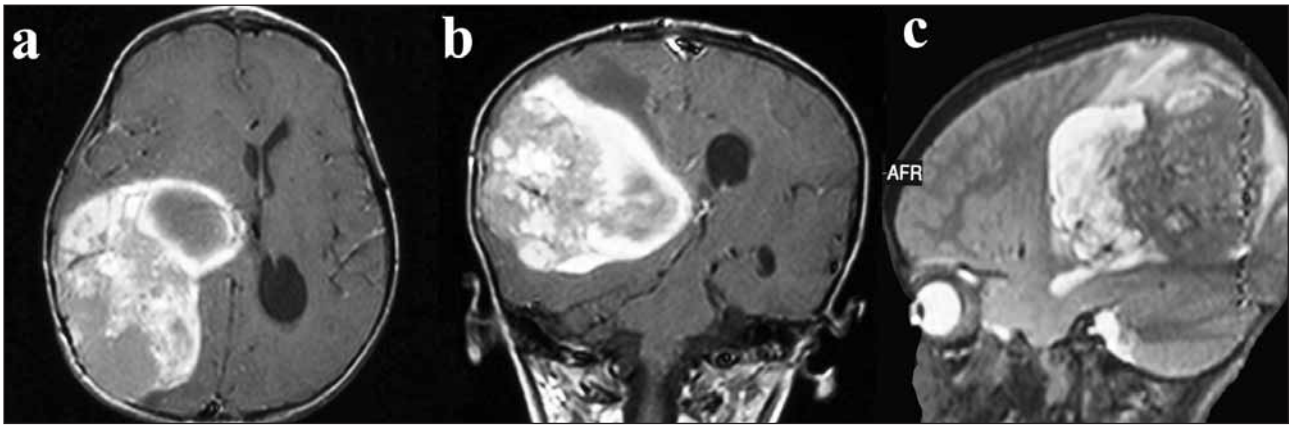

***Figure 1 A-C**: Preoperative axial (**A**), coronal (**B**) and sagittal (**C**) contrast enhanced cranial MRI sections revealed a right parietooccipital heterogenous enhancing cystic mass (dimensions, 8 x 7 x 6,5 cm) and a midline shift of ~ 1,5 cm.*

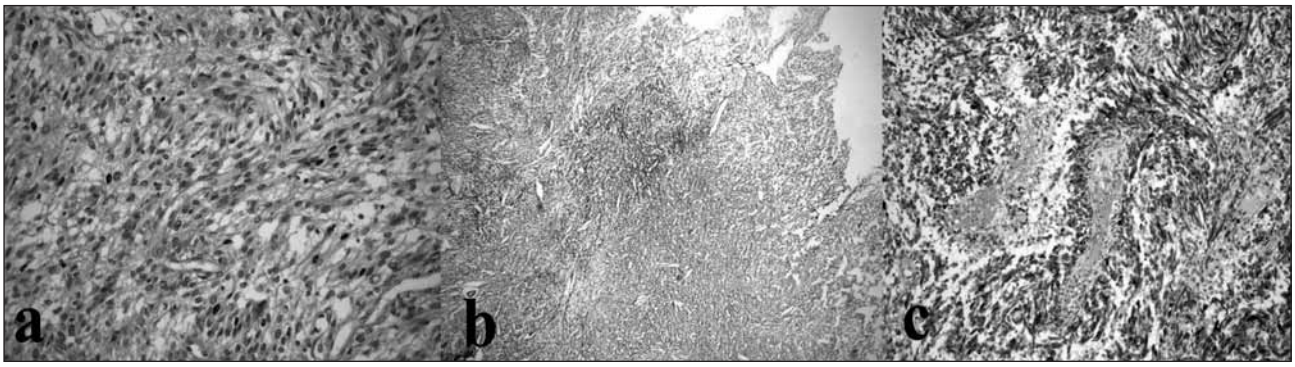

Figure 2A,B,C: ***A****.The tumor was mostly composed of spindle shape astrocytes (H+E x 200) ;* ***B****. It showed conspicuous storiform pattern . Remarkable collagen lay-down in between the astrocytic tumoral cells (reticulin x 100) ;* ***C****. GFAP positive staining confined to the glial component of the tumor (GFAP x l00).*

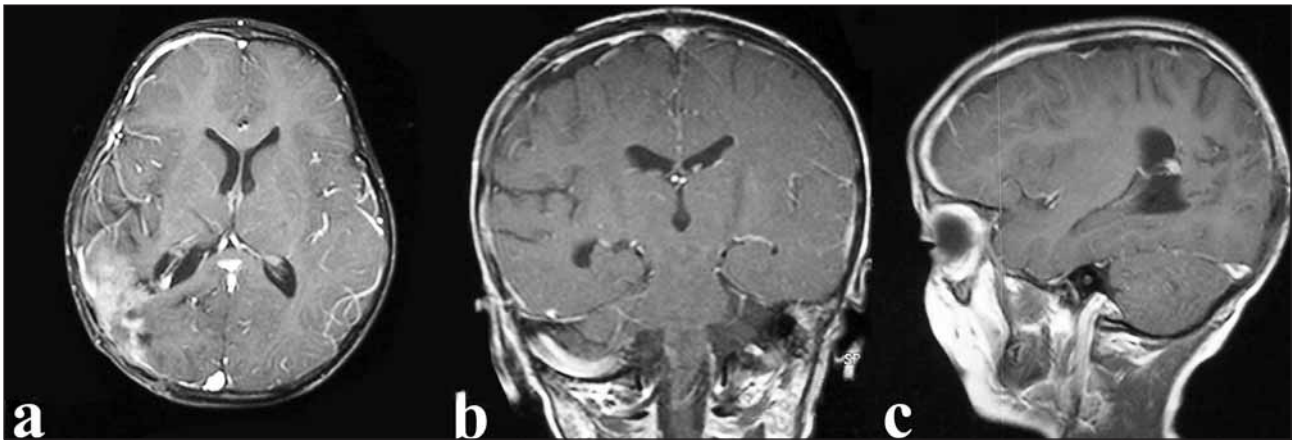

Figure 3A,B,C: *Early postoperative axial (****A****), coronal (****B****) and sagittal (****C****) contrast enhanced cranial MRI sections demonstrate near total removal with postoperative changes.*

Table I. The demographic features of the reported non-infantile DIA cases in the literature.
Abbreviations: FTP: Frontotemporoparietal; T: Temporal; PO: Parietooccipital; SM: Sensory-motor; NM : Not mentioned

Author	Age/Sex	Presenting Sx	Location	Operation	Adjuvant Tx	Follow up
Chacko et al. 1995	7 / F	Seizure	Left FTP	Subtotal	None	24 months No Recurrence
Kurose et al. 2000	9 / M	Seizure & Hemiparesia	Right T	Total	None	6 months No Recurrance
Malucci et al. 2000	3,5 / NM	Hemiparesia	NM	NM	None	No Recurrance
Kato et al. 2004	9 / M	Hemiparesia	Left SM area	Total	None	No Recurrance
Present case	4 / F	Hemiparesia	Right PO	Total	None	11 months No Recurrance

The clinical presentation of DIA is usually within the first 18 months of life (5). Enlarged head circumference, bulging fontanelles and failure to thrive are the most common symptoms and signs. However, these features are absent in noninfantile cases and hemiparesis and seizures predominate the clinical picture. The main symptom was the child's unwillingness to play with her friends in the presented case. She was also reluctant to use her left hand during daily activities. These may be attributed to an attention deficit, unnoticed complex partial seizure and/or left hemiparesis. Moreover, the

duration of the symptoms is usually far shorter in noninfantile cases such as 2 days (2). In the present case, all of the symptoms became marked and established within a week .

Radiologically DIA appears as supratentorial huge mass with broad dural base, having cystic and solid components on CT and MRI. The solid component is contrast enhanced and generally isointense on T1WI and T2WI MRI sections, whereas the cystic component is usually hyperintense in T2WI MRI (3,13). Absence of calcification in DIA has been suggested to aid in differentiating them from the frequently calcified neoplasms of infantile period, although a rare case having dystrophic calcification in the solid component in the absence of dural attachment has also been reported (3). In our case, MRI revealed a right parietooccipital heterogenous enhancing cystic mass (dimensions, 8 x 7 x 6,5 cm) with a midline shift of ~ 1,5 cm.

In the current WHO classification, DIA are presented as Grade I along with desmoplastic infantile gangliogiomas (DIG) (11). DIA share several clinical and histological features with DIG. They are distinguished only by the immunohistochemical demonstration of neuronal cell populations in the latter (8). Pleomorphic xanthoastrocytoma (PXA) also exhibit many similarities with DIA in terms of localization, imaging characteristics and prognosis. Like DIA, PXA's have cortical localization with leptomeningeal involvement, cystic components and good prognosis. However, histopathologically they are more or less easily differentiated from DIA's by the presence of pleomorphic glial cells and lipidized, xanthomatous astrocytes (12). Moreover, PXA's usually occurs in adolescents or young adults (3).

Despite their radiologically aggressive appearance, DIA's are associated with a favorable prognosis even after subtotal resections (7). When complete surgical removal is performed, no recurrence is noted and no further treatment is necessary (13). In a study with a median follow-up of 15.1 years, 6 of 8 patients survived except for 2 intraoperative deaths (11). Thus if the tumor does not allow total removal, adequate decompression rather than radical removal should be the aim in surgery. We were able to perform a radical gross total resection during the operation and did not consider any of the adjuvant therapeutic regimens following the surgery. We have been following the presented case for eleven months and at her last follow-up she had no symptoms and her left hemiparesia was resolved.

There are isolated DIA and DIG cases with metastasis reported in the literature, questioning the benign character of these lesions and the need for chemotherapy and/or radiotherapy as an adjuvant therapy (6,9). However there is still insufficient information available for appropriate adjuvant treatment strategy. This is due to the rarity of these lesions. The patients should be closely followed up and chemotherapy or radiotherapy as an adjuvant treatment should only be considered in case of tumoral progression.

In conclusion, although accepted as a tumor of infancy, DIA can also be encountered in older patients. Careful diagnosis and differentiation with other tumors, particularly malignant astrocytomas is important since the therapeutic strategies may differ. With the increasing awareness of these tumors in the noninfantile period, the current controversies regarding the natural course and optimum treatment methods will be resolved and we believe isolated cases reports like the presented one may contribute to this process.

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