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Chronic subdural haematoma associated with eosinophilic granuloma: a case report in a 13-year-old male child

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Abstract

Subdural haematoma (SDH) is a neurosurgical emergency that can be referred to traumatic or non-traumatic causes. Among causes of SDH, a very rare event is represented by the eosinophilic granuloma (EG), a well-known variety of Langerhans' cell histiocytosis, which seldom is localized inside/under the dura mater. We report herein the case of a 13-year-old boy presenting intracranial hypertension and consequently surgically treated for an imaging revealed SDH. Morpho-histopathological and immunohistochemical (IHC) analyses revealed that SDH was caused by an EG.

Keywords: subdural haematoma, eosinophilic granuloma, Langerhans cell histiocytosis, intracranial hypertension, frontal bone.

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Introduction

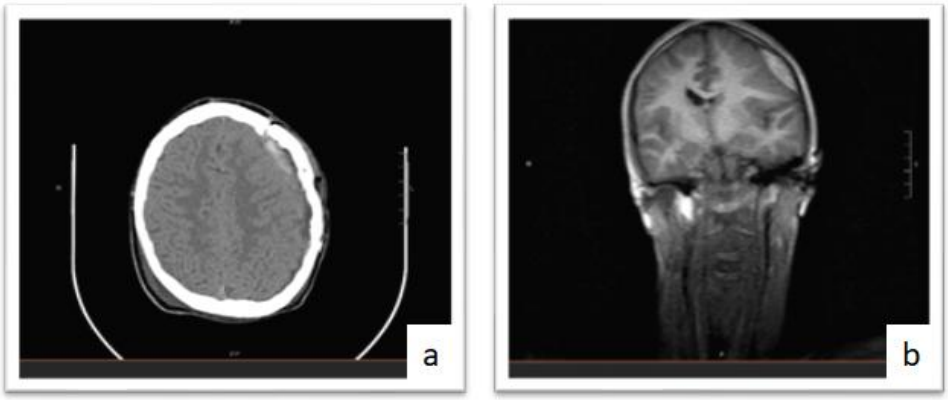
It is well known that the most frequent causes of SDH in children are determined by head injury, congenital malformations, infections, hereditary coagulation disorders, metabolic disorders, tumors and vasculitis (1,2). By contrast, a SDH etiologically- related to EG of the dura mater represents an extremely rare condition. In fact, only few studies have reported an association between epidural haematoma and EG (3); in detail, until now, only three studies have reported cases of EGs growing inside the dural mater, with a secondary invasion of the cerebral tissue and/or the skull (4-6). Generally, EG is characterised by single or multiple skeletal lytic lesions predominantly affecting children, adolescents, or young adults with a good prognosis (7); it may spontaneously regress, or it can be successfully surgically or radiotherapy treated (8). We present herein the case of a 13 years old child suffering from endocranial hypertension due to a SDH associated with an EG.

Case report

A 13 years old male child was admitted to the pediatric first aid department for vomiting and headache, started 8 days before without any history of trauma and haematological investigations

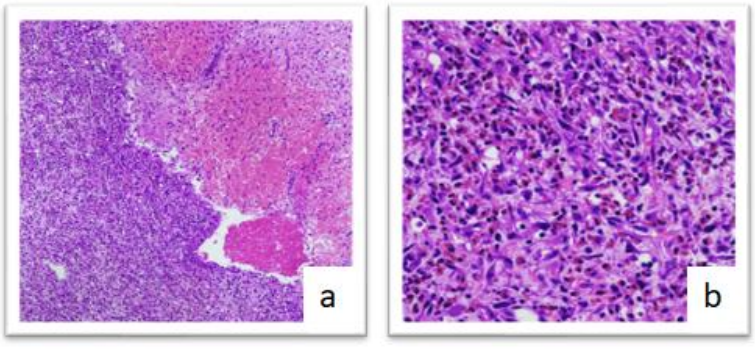
within normal limits. Ophthalmological examination documented fundus papilledema, while Nuclear Magnetic Resonance (NMR) of the brain showed a voluminous extra-axial formation with liquoral intensity, subdural haemorrhage in the left side of the brain associated with light changes in the cerebral morphology and right shift of central encephalic structures (Fig. 1a-b)

Fig. 1. a-b:NMR imagines showing a voluminous extra-axial formation and subdural haemorrhage associated with light changes in the cerebral morphology and right shift of central encephalic structures



Consequently, the subdural haematoma was surgically treated in emergency and the material collected during the procedure was formalin-fixed and transmitted to the histopathology lab. From the corresponding paraffin-embedded tissue blocks, 4-5 micron thick sections were firstly stained by the routine Haematoxylin/Eosin stain. Microscopically, there was an evidence of a fibrotic tissue mixed with blood, showing also an abundant inflammatory infiltration constituted by Langerhans cells (LC), mononuclear histiocyte-like cells with eosinophilic round/oval well defined cytoplasm, oval nuclei with nuclear grooves, also called coffee bean nuclei (Fig.2 a-b); moreover, a huge amount of eosinophilic granulocytes was clearly evident with some giant cells (Fig. 2b). No mitoses neither nuclear/cytoplasmic atypia were encountered.

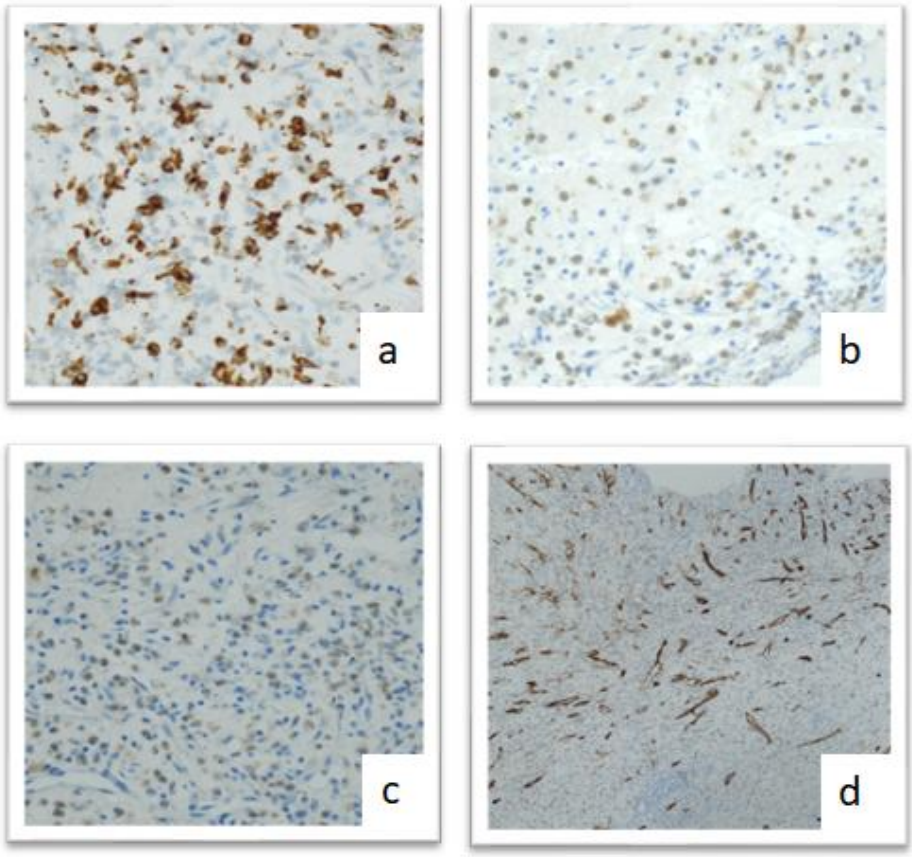
Fig.2. a: Inflammatory infiltration of dural tissue (left) and blood form the haematoma evacuation (right). 2b: detail of the inflammatory infiltration: histiocytes (Langerhans' cells), eosinophilic granulocytes (HE a:10x original magnification; b:40x original magnification



By specific antisera, immunohistochemistry revealed a positive immunostaining for CD68, S-100 and CD1a (Fig.3a-b-c) in Langherans hystiocytic elements as well as in giant cells. Moreover, on serial sections obtained from same tissue blocks, additional immunostainings for CD34, GFAP, EMA and BRAF (V600E) have been performed. By this approach, newly formed intra-lesional

vessels (CD34-positive, Fig. 3d) and the absence of glial elements (GFAP negative) were documented, while some EMA immunoreactive cells showed their external location surrounding the lesion. However, the presence of the protein derived from the genetic mutation of BRAF V600E was not revealed; nevertheless, this mutation of BRAF gene, involved in the signaling pathway RAS-MAPK, has been reported only in 50% of EG cases mainly not located in the skull (10). Finally, on the basis of these morphological grounds, a histopathological diagnosis of EG associated to SDH was finally made.

Fig. 3. Histiocytes showing positive immunoexpression for CD68(a), S100 (b), CD1A (c), intra-lesional vassels showing positive immunoexpression for CD34 (IHC staining, Mayer's haemalum counterstain; a,b,c: 40x original magnification – d: 20x original magnification).



Discussion

The EG is an unifocal rare variety of LC histiocytosis, with a typical histological structure characterized by an incidence of 4-5 cases per million per year in children aged less than 15 years, mainly from 5 to 10 years (11), while adult cases account for 1-2 cases per million per year in adults. The most frequently affected body tissue is represented by the bone, with an osteolytic lesion (axial skeleton, skull, jaw bone, pelvis, rib and long bones) and soft tissues adjacent. Skull and thoracic spine are the elective sites for EG in children, instead jaw and cervical spine in adults (7, 12-16). Other less common localizations are skin, pituitary gland, brain, lung, liver, spleen and gastro-intestinal tract (17-19).

Generally the EG is not associated to SDH, since it is rarely localized in/under the dura mater (4-

6); when it is localized in the skull (more frequently in the frontal bone), it is generally radiologically described as a non-sclerotic punched-out, well-shaped, osteolytic lesion (5,19), which is gradually enlarging and sometimes can involve the underlying dura mater, determining an epidural haematoma (3)

In our case there was no evidence of osteolytic lesion of the skull; nevertheless, a strictly association of the lesion with surrounding EMA reactive meningothelial cells was revealed, suggesting thus the involvement of dura mater instead of the bone. In our opinion, the intradural development of EG should be explained as a consequence of Langherans cells migration in the dural mater caused by a previous flogistic event. Due to a not physiological apoptosis process or to an uncontrolled proliferation, these cells became the physiopathological basis for an EG localized in the subdural space (4,5). However, the hemorrhage mechanism could be explained either with drip phenomena (20) inside the EG or partly with direct compression/mass effect made by the EG over the meningeal vessels (21). In addition, mechanisms such as those above mentioned have been reported in literature in order to explain extradural non traumatic haematoma associated with EG (3).

It is well known that dura mater is a leather-like structure strictly adhering to the periosteum and facing the arachnoid mater; moreover, bridging veins represent additional meningeal structures able to realize little connections through the dura mater. These bridging veins can be damaged if a growing mass, like the EG, stretches their thin walls, releasing blood in the virtual space between dura mater and arachnoid, forming thus the SDH. Of course, the increasing volume of the EG may induce the progressive separation of the meningeal membranes (21); this growing mass, localized in a non-expandable cranial structure, produces compression over its own vessels inside the granuloma and over the bridging veins of the dura mater; by this mechanism, intralesional hemorrhage and rupture of the bridging veins may explain the pathophysiology of the epidural hemorrhage formation. On the other hand, in our case by immunohistochemistry, a fine network of newly formed CD34-reactive vessels has been documented inside the granuloma, probably involved in the haematoma's development. According to these considerations, the mass effect made by the SDH and partly by the volume of the EG itself, clinically revealed evidence of intracranial hypertension, that needed an immediate surgical treatment with craniectomy and evacuation of the haematoma to prevent irreversible damage to noble brain structures.

Conflicts of interest: The authors declare no conflict of interest.

Reference

1. Al-Mousa, A., Altarawneh, M., Alqatawneh, O., Bashir, Z., Al-Dwairy, S., Shtaya, A., (2020). Eosinophilic Granuloma of the Skull Presenting as Non-Traumatic Extradural Haematoma in Children. Int J Gen Med. 24(13), 1229-1234. doi: 10.2147/IJGM.S288512

2. Feldman, K.W., Bethel, R., Shugerman, R.P., Grossman, D.C., Grady, M.S., Ellenbogen, R.G. (2001). The cause of infant and toddler subdural hemorrhage: a prospective study. Pediatrics. 108(3), 636-646. doi: 10.1542/peds.108.3.636.

3. Al-Mousa, A., Altarawneh, M., Alqatawneh, O., Bashir, Z., Al-Dwairy, S., Shtaya, A. (2020) Eosinophilic

Granuloma of the Skull Presenting as Non-Traumatic Extradural Haematoma in Children. Int J Gen Med. 24(13),1229-1234. doi: 10.2147/IJGM.S288512.
4. Caroli, E., Ferrante, L. (2007) Intradural eosinophilic granuloma with intraparenchymal invasion: a new growth pattern. ZentralblNeurochir. 68(2), 79-82. doi: 10.1055/s-2007-980172.
5. Mardi, K., Thakur, R.C., Negi, L. (2017) Intradural Eosinophilic Granuloma Invading Skull: Case Report and Review of the Literature. Asian Journal of Neurosurgery. 12(4), 698-700. doi: 10.4103/ajns.AJNS_47_15.
6. Kazanci, B., Tehli, O., Kazanci, A., Guclu, B. (2014) Intradural Langerhans' cell histiocytosis invading skull: Case report and review of the literature. J Neurol Sci. 31, 609-613.
7. Sunder, G., Mukesh, P., Deepika P. (2020). Intradural Langerhans' Cell Histiocytosis Invading Frontal Bone of Skull: A Case Report with A Brief Review. Annals of Clinical and Medical Case Reports. 05. 10.47829/ACMCR.2020.5301.
8. Dukmak, O.N., Abualia, S. M. S., Meqbil, Y. J. I., Emar, M., Basal, S.I, Itaidek, S. (2022) Eosinophilic granuloma affecting the parietal bone of the skull: A case report and literature review. International Journal of Surgery Case Reports. 96(107371), 2210-2612. doi: 10.1016/j.ijscr.2022.107371.
9. Kaul, R., Gupta, N., Gupta, S., Gupta, M. (2009). Eosinophilic granuloma of skull bone. J Cytol. 26(4). 156-157. doi: 10.4103/0970-9371.62188.
10. Kumar, N., Sayed, S., Vinayak, S. (2011). Diagnosis of Langerhans cell histiocytosis on fine needle aspiration cytology: a case report and review of the cytology literature. Patholog Res Int. 20. doi: 10.4061/2011/439518.
11. Tran, G., Huynh, T.N., Paller, A.S. (2018). Langerhans cell histiocytosis: A neoplastic disorder driven by Ras-ERK pathway mutations. J Am AcadDermatol. 78(3), 579-590.e4. doi: 10.1016/j.jaad.2017.09.022.
12. Allen, C.E., Ladisch, S., McClain, K.L. (2015). How I treat Langerhans cell histiocytosis. Blood. 126(1). 26-35. doi: 10.1182/blood-2014-12-569301.
13.Islinger, R.B., Kuklo, T.R., Owens, B.D., Horan, P.J., Choma, T.J., Murphey, M.D., Temple, H.T., (2000) Langerhans' cell histiocytosis in patients older than 21 years. ClinOrthopRelat Res. (379), 231-235. doi: 10.1097/00003086-200010000-00027.
14. Cochrane, L.A., Prince, M., Clarke, K., (2003) Langerhans' cell histiocytosis in the paediatric population: presentation and treatment of head and neck manifestations. J Otolaryngol. 32(1), 33-37. doi: 10.2310/7070.2003.35266.
15. Sherwani, R.K., Akhtar, K., Qadri, S., Ray, P.S. (2014). Eosinophilic granuloma of the mandible: a diagnostic dilemma. BMJ Case Rep. 2014(3);2014. doi: 10.1136/bcr-2013-200274.
16. Bajracharya, B., Poudel, P., Bajracharya, D., Bhattacharyya, S., Shakya, P. (2018) Eosinophilic Granuloma of Mandible: A Diagnostic Challenge. Kathmandu Univ Med J (KUMJ). 16(62), 201-203. PMID: 30636766.
17. Prasad, G.L., Divya, S. (2019) Eosinophilic Granuloma of the Cervical Spine in Audults: A Review. World Neurosurg. 125, 301-311. doi: 10.1016/j.wneu.2019.01.230.
18. Öğrenci, A., Batçık, O.E., Ekşi, M.Ş., Koban, O. (2016) Pandora's box: eosinophilic granuloma at the cerebellopontine angle-should we open it? Childs Nerv Syst. 32(8), 1513-1516. doi: 10.1007/s00381-015-2982-1.
19. Haupt, R., Minkov, M., Astigarraga, I., Schäfer, E., Nanduri, V., Jubran, R., Egeler, R.M., Janka, G., Micic, D., Rodriguez-Galindo, C., Van Gool, S., Visser, J., Weitzman, S., Donadieu, J., Euro Histio Network. (2013) Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 60(2), 175-184. doi: 10.1002/pbc.24367.
20. Jha, S.K., De Jesus, O. (2022) Eosinophilic Granuloma. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–. PMID: 32644464.
21 Sadashiva, N., Baruah, S., Rao, S., Mahadevan, A., Rao, K.N., Vazhayil, V., Somanna, S. (2017) Eosinophilic Granuloma of Skull with Fluid Level and Epidural Hematoma: A Case Report and Review of the Literature. PediatrNeurosurg. 52(1), 41-45. doi: 10.1159/000448047.
22. Bakhaidar, M.G., Alghamdi, F.A., Baeesa, S.S. (2016) Spontaneous extradural hemorrhage due to Langerhans cell histiocytosis of the skull in a child: a rare presentation. J PediatrNeurosci. 11(1), 52–55. doi: 10.4103/1817-1745.181248

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