CASE REPORT

Intracranial angiomatoid fibrous histiocytoma with Hodgkin lymphoma

Gunness Vimal Raj Nitish, MD[1], Munoz David G, MD[2], Mikhalkova Agdaliya, MD[3], González-López Pablo, MD, PhD[4], Alshafai Nabeel, MD[5], Spears Julian, MD[1]

[1]Neurosurgery Department, St. Michael's Hospital, Toronto, Ontario, Canada, [2]Department of Laboratory Medicine St. Michael's Hospital, Toronto, Ontario, Canada, [3]Neurosurgery Department, Academic Teaching Hospital of Feldkirch, Feldkirch, Austria, [4]Neurosurgery Service, Alicante University General Hospital, Alicante Institute for Health and Biomedical Research (ISABIAL-FISABIO Foundation), Avda. Pintor Baeza sn, Alicante, Spain, [5]Neurosurgery Department, Royal Commission Hospital, Al Lulu Road, Fanateer, Al Jubail, Saudi Arabia

SUMMARY

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumour of uncertain differentiation and low metastatic potential, which occurs predominantly in children and young adults. It occurs mostly within the extremities, trunk, head and neck. We report the case of a 32-year-old female that was operated in our hospital in 2016 and twice in 2017. The patient had headaches and neck pain initially in 2016. We discuss the radiographic and histologic features initially found and the findings that ultimately led to the diagnosis of AFH. The patient had a past history of Hodgkin lymphoma.

INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumour with intermediate malignant potential.[1] In 1976, Gonzalez-Vitale et al., described a similar pathology after a patient received a heavy dose of radiation in the sella turcica region. The first time the term AFH was used was in 1979, when Enziger described a series of 41 unusual examples.[2]

AFH is rare, accounting for approximately only 0.3% of all soft tissue tumours, although given its histologic similarity to a variety of other neoplasms it is likely that it has been previously underdiagnosed and subsumed under a variety of other neoplastic categories, including vascular, fibrohistiocytic, and myofibroblastic types.

Larger studies of AFH have shown a favourable prognosis with gross-total resection, leading to its identification as a separate entity from malignant fibrous histiocytoma. As more specific markers are becoming available, this rare tumour is being identified in more locations than previously noted.

Five reports of AFH have been published for intracranial lesions and four were primary lesions and one was considered a metastasis.[3]

In this report, we describe this rare intracranial tumour in a patient, who previously had Hodgkin lymphoma. She was subsequently operated on for the second time due to the recurrence of the tumour and received a cystoperitoneal shunt for an intracerebral cyst afterwards.

CASE REPORT

A 32-year-old female of Caucasian origin was admitted to our clinic on September 2016 with severe headache and neck pain. She was seen first in another hospital, where papilledema was found during eye examination. The patient had a computed tomography (CT) head followed by magnetic resonance imaging (MRI) (Fig 1), which showed a sharply demarcated lobulated heterogenous, intense enhancing mass with an extension into right middle temporal gyrus predominantly in the atrium of the right lateral ventricle.

Neurological status at admission: GCS 15, no cranial nerve deficiency.

The past medical history of the patient included cholecystectomy and classical nodular sclerosing Hodgkin lymphoma in 2007 which was stage IIA. She was treated with ABVD (chemotherapy regimen) biweekly for six months. She did not have any radiotherapy.

The patient was operated through a right temporal craniotomy with intraoperative neuronavigation, and she was discharged two days after the surgery. A left upper quadrantanopia was found at the postoperative eye examination.

Intraoperatively the lesion looked well circumscribed, solid and resembled more a meningioma than an intra-axial tumour.

Histopathological results came out as myxoid lesion, consistent with angiomatoid fibrous histiocytoma. Microscopically the lesion was composed of cells with round, ovoid, tapered, serpentine and pleomorphic nuclei present in eosinophilic or vacuolated cytoplasm growing in sheets and whorls in a myxoid stroma (Fig 2) characteristic of angiomatoid fibrous histiocytoma. Ki67=5%. The neuropathologist performed a fusion panel.

The patient was followed up by regular MRIs and she refused any oncological treatment.

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Corresponding Author: Vimal Raj Nitish Gunness
Email: nitish57@yahoo.com

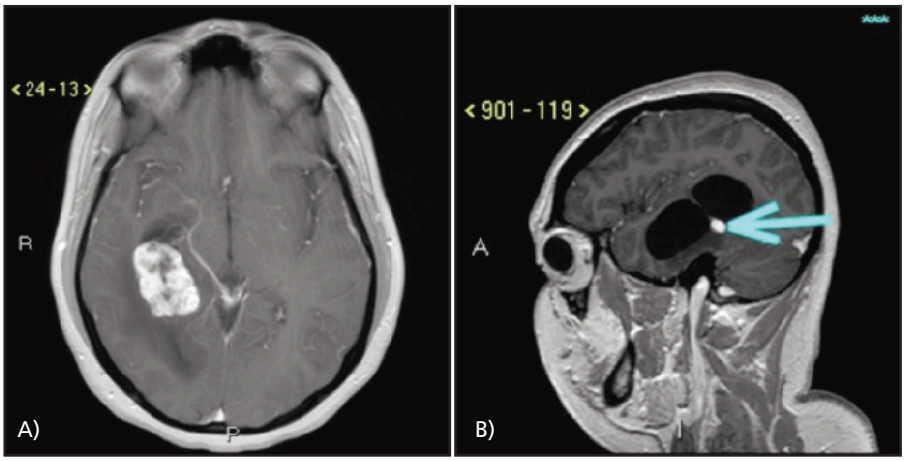

Fig. 1: A) Magnetic Resonance Imaging (MRI) - axial gadolinium shows heterogenous, hyperintense, cauliflower appearance. B) MRI - sagittal T1 with gadolinium showing a hyperdense nodule.

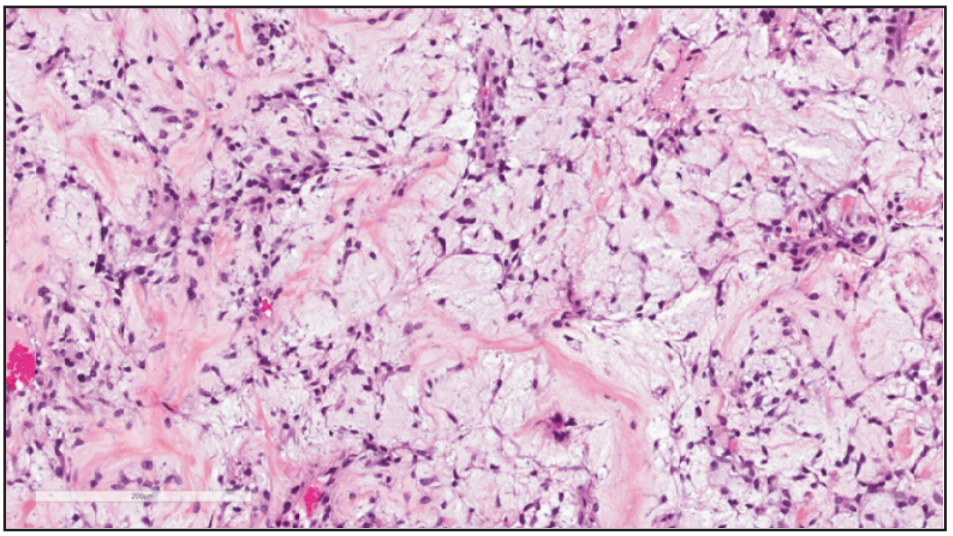

Fig. 2: Stain is HPS (Hematoxylin-Floxine-Saffron).

In September 2017 on one of her follow-up MRIs we saw a recurrence measuring almost 1cm. In addition, there was a trapped ventricle on the level of the right temporal horn.

She was seen at our neurosurgery clinic and the treatment options were discussed including radiosurgery, conventional radiation and redo surgery. She wished to proceed with the second surgery.

On 19 October 2017, the patient was operated again. Intraoperatively we identified the gross abnormality under the microscope. It looked like a small, fairly well-defined nodule embedded in the pia matter.

Postoperatively a focal seizure to the right eye and arm occurred and lasted for approximatively 30 seconds. Consequent head CT was done and revealed no postoperative bleeding. The postoperative MRI showed the expected postoperative changes and no definite evidence of residual enhancing tumour.

The histopathological report came as angiomatoid fibrous histiocytoma (Fig 2) with similar morphology as the previous lesion.

Two weeks after being discharged from her second surgery, the patient presented to the Emergency Department with a history of neck pain and right sided headaches for one week. She did not have any fever, chills or nausea to suggest a source of infection.

The CT and MRI scans showed a right sided temporal horn enlargement with an adjacent fluid filled cyst. We suggested the patient to undergo a CSF diversion procedure in order to relieve the increased intracranial pressure and alleviate her symptoms. On 9.11.2017, she underwent the placement of a cystoperitoneal shunt. The postoperative head CT showed good shunt placement, and the cyst was decreased in size.

Her headaches improved substantially after the operation. The patient was ambulating safely and independently and was discharged home one day after the surgery.

On 4 December 2017, she did not complain of any new symptoms and the headaches disappeared. The patient was followed up with a new MRI in April 2018 and September 2018 which did not show any relapse.

DISCUSSION

AFH is a rare soft tissue neoplasm of intermediate (rarely metastasising) biologic potential and uncertain differentiation, which predominantly arises superficially in the deep dermis and subcutis of the extremities of children and young adults.[4]

There is little documentation of similar lesions in the literature in the past, perhaps because they fell under descriptions of other entities such as sclerosing haemangioma, dermatofibroma, and other lesions classified as fibrous histiocytoma.

AFH accounts for approximately only 0.3% of all soft tissue tumours,[4] although given its histologic similarity to a variety of other neoplasms it is likely that it has previously been underdiagnosed and subsumed under a variety of other neoplastic categories, including vascular, fibrohistiocytic, and myofibroblastic types. Patients present with AFH in the first three decades of life, although the age distribution is wide, it can occur in infants (including congenital cases), and adults in the eighth and ninth decades. There is no significant sex predilection.

Reports of the AFH occurrence as a second neoplasm in other malignancies are rare and include a supraclavicular AFH occurring in a 27-year-old man 16 months after chemotherapy for disseminated testicular cancer, an inguinal mass in a child with stage III retroperitoneal neuroblastoma and in an 18-year-old woman with a 4-year history of treatment for Hodgkin lymphoma.5 It is unclear whether these tumours may have occurred secondarily to the treatment, although it appears more likely that they were incidental.

Less than 5% of AFHs have been reported to metastasize sometimes after multiple recurrences, predominantly to regional lymph nodes and rarely to the lungs, liver, or brain. There is no published data on how AFH evolves on long term intracranially. Grossly, AFHs are firm, multinodular and multicystic, haemorrhagic masses, with cut surfaces varying from greyish-yellow to white. They are usually small, ranging from 2 to 4 cm, although they can reach 10cm (median, 2.5cm).[3]

Angiomatoid fibrous histiocytoma is associated with the following three characteristic translocations: t(2:22)(q33:q12) (forming the EWSR1-CREB1 fusion gene), t(12:22)-(q13:q12) (forming the EWSR1-ATF1 fusion gene) and t(12:16)(q13:p11) (resulting in the FUS-ATF1 fusion gene). EWSR1 and FUS are multifunctional proteins belonging to the FET (previously TET) family of RNA-binding proteins, which are implicated in central cellular mechanisms such as the regulation of gene expression, maintenance of genomic integrity, and processing of messenger RNA/microRNA.

Our case, the 5th primary intracranial AFH lesion, illustrates several unique features. The patient had Hodgkin lymphoma 10 years earlier which was treated by chemotherapy. This is the first case of an intracranial AFH in a patient with the previous history of Hodgkin lymphoma. Although Pettinato G et al., reported a patient with Hodgkin lymphoma, it was not clear where the AFH lesion was situated.5 Further, it came as usual cyst formation at the site of the previous tumour. All these features make this case unique.

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