



Case Report

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Challenging Diagnosis of Cerebellum Hemangioendothelioma in Presence of Tb: A Case Report



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Summary

hemangioendotheliomas are rare malignant, invasive tumors and can be misdiagnosed or over looked by another symptoms of primary disease There is no clear guideline to guide treatment. In this case report, we report case of CNS TB which masked diagnosis of Hemangioendothelioma of the brain. The patient was treated for CNS TB for 18 months with excellent response for all lesions in the brain and extracranially except for one lesion in cerebellum which turned out to be hemangioendothelioma.

Introduction

The term epithelioid hemangioendothelioma (EHE) was described in 1982 for the first time by Weiss and Enzinger [1]. It is a rare vascular tumor caused by abnormal growth of blood vessels. Although the exact underlying cause for the abnormal growth is unknown, it occurs mainly in adults [2]. Epithelioid hemangioendothelioma mainly involves skin and soft tissues compared to deep organs [1].

World Health Organization WHO classification describes Epithelioid Hemangioendothelioma as it does contain an admixture of histologically distinct components [3]. We describe in this case report the process of diagnosis of the first cerebellum hemangioendothelioma in presence of extrapyramidal Tuberculosis (TB) reported in Saudi Arabia.

Case Report

A patient consent and IRB approval was obtained before starting to collect data of this interesting case report.

The case report was about a 52-year-old male gentleman presented in 2018 with progressive right upper limb weakness and weight loss about 20Kg. Investigations which included MRI brain revealed multiple supra and infra-tentorial ring lesions in brain (Figure 6). CT chest, abdomen and pelvis showed wide

spread bony lytic lesions and mediastinal lymph nodes. A series of culture and biopsy was taken from left parietal lesion; it was negative for TB. There was no malignancy seen. Bronchoscopy lavage was positive for TB. Patient was started on anti-TB medication and was discharged home with follow-up in neurosurgery and infectious disease outpatient clinic follow-up.

Patient was followed up with serial brain MRI. MRI of brain in 2019 showed mixed response. Repeat MRI brain end of 2019 showed overall regression in numbers and sizes. Patient was followed up with serial brain MRI until 2021; right cerebellar lesions started to show disease progression while other lesions remain stable.

Patient then went to another hospital and had debulking surgery. Histopathology reported as differentiated carcinoma. A slide review in our hospital reported as hemangioendothelioma. After this the patient lost follow-up for 4 months then presented again to our hospital with worsening headache. MRI showed cystic lesion which increases in size. Patient underwent second debulking surgery at our hospital for the posterior fossa lesion. The pathology sample was consisting of fragments of white and grey, friable, hemorrhagic tissue aggregate 4.0 x 3.0 x 2.0cm. Microscopically, it showed vascular neoplasm consistent with hemangioendothelioma (figure 1 & 2). Immunohistochemical examination for FLI-1 and CD 31 stain shows strong and diffuse

nuclear staining in neoplastic cells (figure 3 & 4). Tumor cells are focal positive for pan-CK EMA, CD34, S100, GFAP, SMA and PR immunostains are negative (Figure 5). Pathology confirmed recurrence of hemangioendothelioma. Patient was referred to

radiotherapy. He was referred to radiotherapy. Patient underwent radiotherapy 54gy in 30 fractions. Patient tolerated radiotherapy very well. MRI scan after 3 months showed regression of cystic lesion in the right cerebellum (Figure 6).

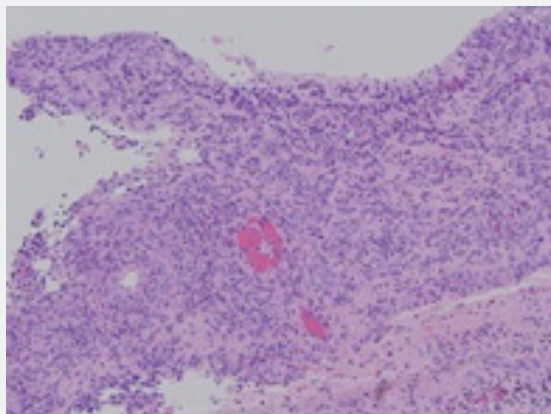


Figure 1: Microscopic examination (H&E) at high power showing spindle cell neoplasm, plump eosinophilic cytoplasm vesicular nuclei with focal prominent nucleoli.

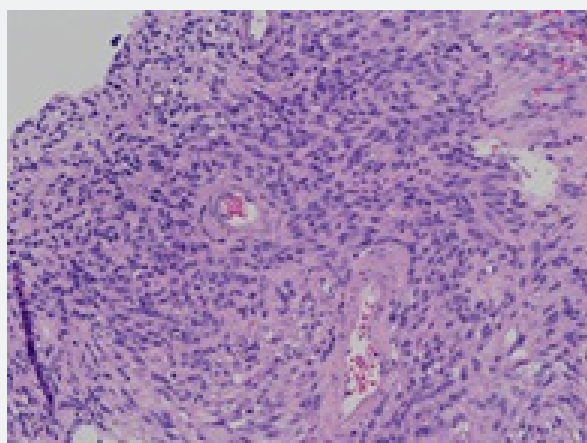


Figure 2: Histologic examination (H&E) at low power show neoplastic cells invading brain tissue.

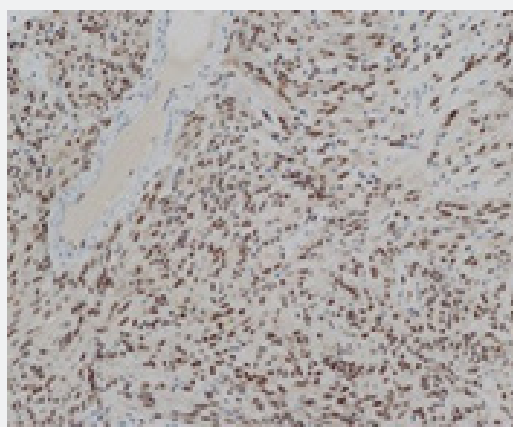


Figure 3: Immunohistochemical examination for FLI-1 stain shows strong and diffuse nuclear staining in neoplastic cells.

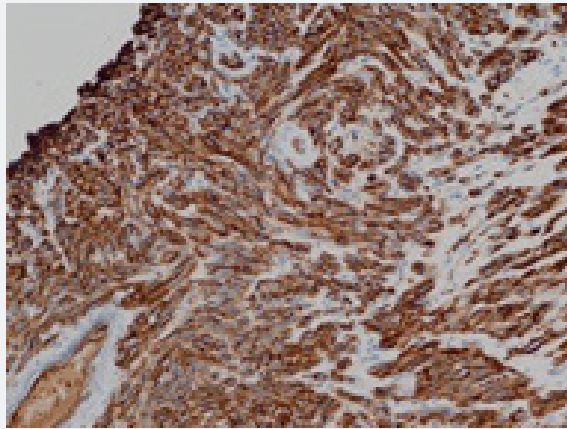


Figure 4: Immunohistochemical examination for CD31 shows strong and diffuse membranous staining in neoplastic cells.

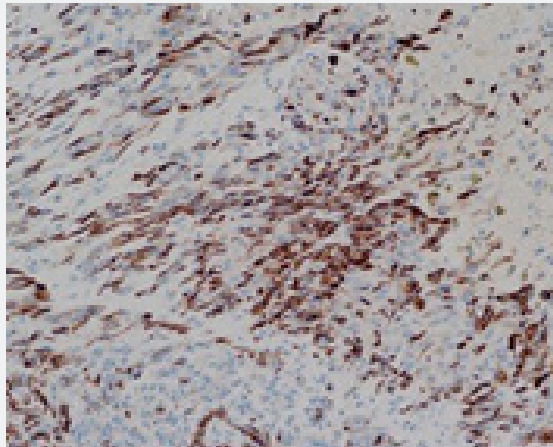


Figure 5: Tumor cells are focal positive for pan-CK EMA, CD34, S100, GFAP, SMA and PR immunostains are negative.



Figure 6: MRI images of brain showing multiple rounded lesions in cerebral hemisphere and cerebellum bilaterally. The lesion on **the right** cerebellum was Hemangioendothelioma.

Discussion

Hemangioendothelioma is very rare disease and the most common location was reported on soft tissue, lung, liver and bone. It is rarely present intracranially [4]. CNS TB is much more common compared to Hemangioendothelioma in cerebellum.

According to the Canadian guidelines the diagnosis of non respiratory TB, biopsy of affected organ should be sent for AFB and TB culture. Respiratory TB should be investigated. Hemangioendothelioma is difficult to diagnose with radiological imaging due to its rarity. Biopsy is required for establish the diagnosis [5].

In our case, there was coexisting dual pathology, TB and EHE. Both have different management strategy with different outcomes. The clinical presentation of limb weakness and weight loss which make it more complicated to differentiate between 2 pathology. Radiological findings of MRI in the form of T1 hyperintense lesions was reported in both brain T.B [6] and EHE [7]. A recent report of a case of spinal hemangioendothelioma which was initially misdiagnosed as a spinal TB by the diagnostic MRI ultimately , tissue diagnosis was the only way to reach the accurate diagnosis in this complicated case [8]. Linalidomide has been reported to have a good disease control even with CNS EHE [9].

In our case the challenging diagnosis is the simultaneous occurrence of two completely distinct pathologies; TB and EHE. Clinical presentation in the form of (limb weakness and weight loss) loss was indistinguishable and Also radiological findings in the baseline MRI in the form of T1 hyperintense lesions was reported in both brain T.B and EHE, Xie and his colleagues recently reported a case of spinal hemangioendothelioma which was initially misdiagnosed as a spinal TB by the diagnostic MRI, ultimately , tissue diagnosis was the only way to reach the accurate diagnosis in this complicated case.

Conclusion

We report first case of intracranial TB coexisting with EHE. Tissue diagnosis remain the gold standard to reach final diagnosis of EHE. EHE can be misdiagnosed for another disease such TB.

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