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A Rare Case of Cerebral Hemangioblastoma Mimicking as Anterior Choroid Plexus Cyst



حالة نادرة تورم أرومي وعائلي في الدماغ، يشبه كيس صغيرة مشيمية أمامية.

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Abstract

المستخلص

A choroid plexus cyst may be present throughout the ventricular system of the brain. A hemangioblastoma is generally found in patients with genetic diseases like Von Hippel-Lindau (VHL) syndrome disease, but it can also be found sporadically in the spinal cord, retina and in rare cases in the cerebrum.

An eighteen-year-old male subject was referred for autopsy in Kolkata police morgue on 27/02/2017. Two hours before, the patient had been brought to the hospital for severe headache; but before any diagnosis could be made, he collapsed and died. On autopsy, a 4cm fluid filled cyst was found in the anterior portion of third ventricle and sent for histopathological examination. HPE findings were consistent with a hemangioblastoma, which made this case extremely rare.

Sudden death due to increased intracranial tension cases should be properly examined for any neuroepithelial tumours like a hemangioblastoma.

قد يكون كيس الضفيرة المشيمية موجوداً في جميع أنحاء البطين في المخ. وعادة ما تصادف الأورام الأرومية الوعائية مصاحبة لأمراض وراثية مثل مرض متلازمة (VHL Von Hippel-Lindau)، ولكن يمكن أيضاً أن توجد بشكل متقطع في الحبل الشوكي أو شبكية العين ونادراً ما توجد في المخ.

تم إحالة أحد الذكور البالغ من العمر ثمانية عشر عاماً للتشريح في مشرحة شرطة كلكتا بتاريخ 27/02/2017 م. حيث أنه قبل ساعتين كان قد أحضر المريض إلى المستشفى بسبب وجود صداع شديد لديه، ولكن قبل إجراء أي تشخيص له انهار ومات. وأثناء تشريح الجثة، شوهد كيس مملوء بسائل حجمه حوالي 4 سم، وجد في الجزء الأمامي من التجويف الثالث وقد تم إرساله للفحص النسيجي. كانت نتائج الفحص النسيجي (HPE) متسقة مع أن ذلك الكيس هو ورم وعائلي، ما جعل هذه الحالة تعد حالة نادرة للغاية.

وعليه فإنه يجب فحص حالة الموت المفاجئ الناجم عن زيادة حالات التوتر داخل الجمجمة بشكل صحيح وذلك تحسباً لوجود أي أورام عصبية ظاهرية مثل الورم الأرومي الوعائلي الدموي.

Keywords: Forensic Science, Forensic Medicine, Autopsy, Cerebral Hemangioblastoma, Lateral Ventricle.

الكلمات المفتاحية: علوم الأدلة الجنائية، الطب الشرعي، التشريح، الورم الأرومي الوعائلي الدماغ، البطين الجانبي.



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1. Introduction

A choroid plexus cyst may be present throughout the ventricular system of the brain. In 1% of pregnancies it affects fetuses, but in adults it is rare and asymptomatic, generally being found incidentally during autopsy [1]. Hemangioblastomas of the central nervous system are said to constitute 1.1 to 2.4 percent of all the intracranial neoplasms. Almost all occur in the cerebellum, occasionally being found in the pons, medulla, and spinal cord. However, cerebral hemangioblastomas are extremely rare [2]. In the case presented in this paper, a lesion was present in the anterior part of the lateral ventricle (shown in the video link). Histopathologically, it was hemangioblastoma, which is extremely rare.

2. Case Report

An eighteen-year Indian male subject was referred to the Kolkata police morgue on 27/02/2019 from a primary health centre in West Bengal for autopsy to determine cause of death. Information given by police revealed that the patient complained of a headache the previous night and was taken to the hospital. On the day of admission, after 2 hours, the patient collapsed and was declared dead by the attending medical officer before he could make any proper diagnosis. According to the treatment file, the patient was treated with IV Fluids, injection mannitol and analgesic. The patient's relatives said that he had occasional headaches which were not so severe and was treated conservatively; however, the last headache was intense.

3. Materials and Methods

All the standard autopsy instruments were used to perform the autopsy. Autopsy was performed on the dead subject following modified Ghon's technique.

3.1 External Examination

The corpse of a moderately built (63.58 Kg) subject with rigor mortis all over the body was presented. Pupils did not react to light bilaterally; livor mortis was present over the dependent part of body, except at the pressure points. No external injury was found, even after careful examination under hand lens.

3.2 Internal Examination

The body was opened with a conventional "I" incision and different organ systems were carefully examined. None of the internal organs showed any hemorrhage, cyst or injuries with naked eye examination.

On opening of the cranial cavity, the brain was found to be edematous and heavy (1,698 g) after it was taken out. On dissection of the corpus callosum, one cyst filled with clear fluid was found in the anterior portion of the lateral ventricle (Ref. the video link). The diameter of the cyst was 2.1 cm, and it was attached with a red colored stalk whose length was 7 cm. The cyst and the stalk were carefully dissected from the ventricular wall and kept in 10% formol saline solution for histopathological examination. Ventricular walls were found to be thinned out and the cavity was enlarged, which suggested increased pressure within the ventricles. No other hemorrhage or injuries were found in brain.

3.3 Histology

The tissue obtained was processed in an automated tissue processor as per standard methods and finally stained with Hematoxylin and Eosin stain.

4. Results

Sections from the cystic specimen showed the vascular component consisting of small capillaries with a single



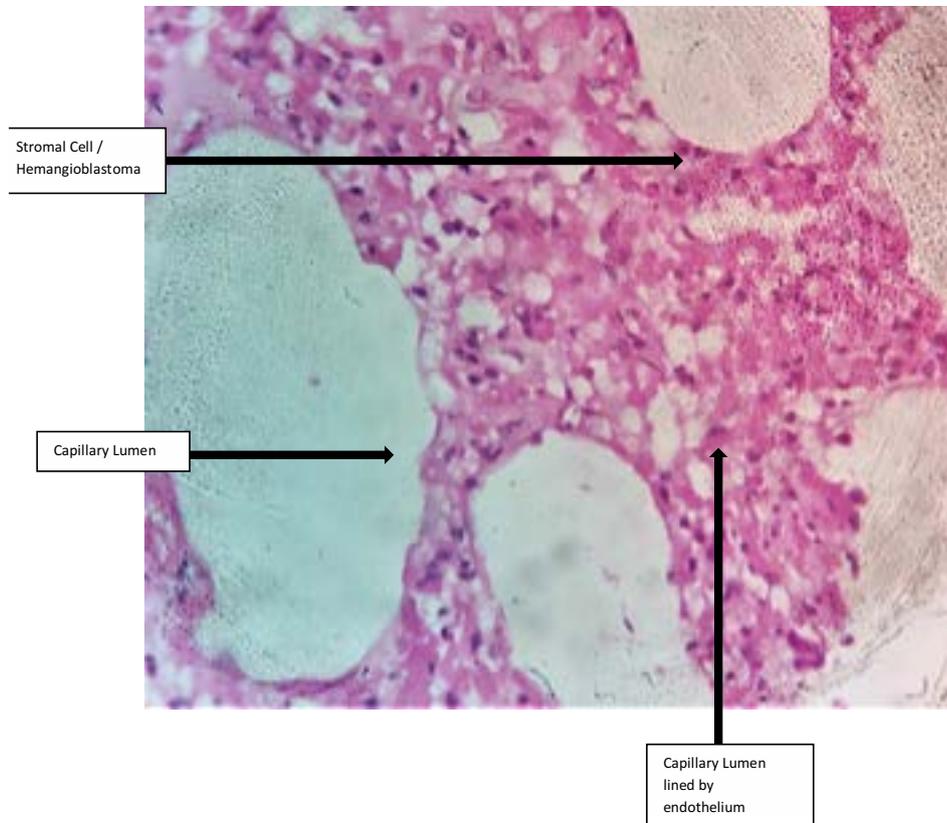


Figure 1- Capillaries of different dimension and lined by endothelium, stromal cells looking as empty spaces in between (H&E Stain).

layer of plump, uniform endothelial cell. The cellular component consisted of large and vacuolated stromal cells, features of a hemangioblastoma [3]. The histological findings in this case was exactly the same as described [3] (Figure-1). Choroidal lining was seen outside the cystic structure. Other parts of the given brain tissue were unremarkable. Sections from lung, liver, kidney and pancreas were all normal.

5. Discussion

In 1928, Cushing and Bailey introduced the term hemangioblastoma [4]. It refers to a benign vascular neoplasm that arises from almost exclusively the central nervous system. Predominant involvement of the cerebellum and spinal cord was noted, but true incidence of tumour was not discovered till increased availability of magnetic

resonance imaging. The aetiology of a hemangioblastoma is obscure, but its presence in various clinical syndromes may suggest an underlying genetic abnormality. The genetic hallmark of a hemangioblastoma is the loss of function of the Von-Hippel-Lindau (VHL) tumour suppressor gene [5].

Upon gross examination, hemangioblastomas are usually cherry red. They may include a cyst that contains a clear fluid, but solid tumours are as common as cystic ones. The tumour usually grows inside the parenchyma of the cerebellum, brain stem, or spinal cord; it is attached to pia matter and gets its rich vascular supply from pial vessels [6]. Depending upon the position of a hemangioblastoma, symptoms vary from cerebellar dysfunction like balance, muscle tone equilibrium and difficulty in walking, standing or moving in a coordinated manner to increased intracra-

nial pressure and hydrocephalus. Symptoms of a ruptured hemangioblastoma are often dramatic as increased pressure causes compression of brain structure [7]. In many cases, symptoms caused by the growth of the neoplasm itself may be an indication for surgical intervention. In others, symptomatic obstruction of a cerebrospinal fluid pathway may necessitate the operation. Important differential diagnosis is a Choroidal plexus cyst, which is the most common neuroepithelial cyst. Typically, they are small and asymptomatic but are rarely large (2-8 cm) [8,9]. Primary brain tumors which cause sudden death and are found accidentally during autopsy are glioblastoma multiforme, anaplastic astrocytoma, oligodendroglioma, medulloblastoma, lymphoma, teratoma and pituitary adenoma [10,11].

6. Conclusion

Sudden death due to increased intracranial pressure cases should be properly examined for any neuroepithelial tumors. The most likely cause of death in the case presented above was also due to the same reason which in turn resulted from acute hydrocephalus. Histopathology of the tumors is of utmost importance in conformation of diagnosis. Death in such cases can only be avoided if we have a high degree of suspicion for such ailments.

The video is available in the following link - <https://www.youtube.com/watch?v=26LQ01U190Y>

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Conflict of Interests

Nil

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