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A Cerebellar Adenocarcinoma misdiagnosed as a Haemangioblastoma.

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Abstract

A patient with a presumed cerebellar haemangioblastoma was referred for stereotactic radiosurgery after initially undergoing an unsuccessful attempt at surgical excision. The lesion subsequently proved to be a cystic adenocarcinoma

Keywords: cerebellum, haemangioblastoma, cystic adenocarcinoma, stereotactic radiosurgery, histology

Case Presentation

The patient presented (in October 2017) with a chronic history of altered sensation in the left side of her face and dizziness (more than 6 months) and more recent ‘clumsiness’ of her left leg(weeks). She was a known hypertensive, drank alcohol regularly by her own admission and had smoked 25 cigarettes a day for 50 years. There were no constitutional symptoms. There was neither a family history of cancer nor neurocutaneous syndrome(s). The patient’s

fingers exhibited marked osteoarthritic changes of the distal interphalangeal joints (Heberden's nodes) but no finger clubbing.

On clinical examination broad-based ataxic gait and mildly staccato speech with no gross appendicular cerebellar signs was noted. MRI of the brain demonstrated a left cerebellar cyst with a minute area of nodular enhancement (the wall of the cyst did not enhance and there was no associated edema). CT scan of the chest demonstrated a heavily calcified lesion in the upper lobe, with no lymphadenopathy, thought to be benign by both the attending cardiothoracic surgeon and reporting radiologist. CT scan of the abdomen did not reveal any mass lesions. Blood parameters and fundal eye examinations were normal.

The patient initially refused surgery but consented thereto 6 months later because of progressive incapacitating ataxia. The MRI appearance of the lesion was unchanged at this time and a diagnosis of sporadic haemangioblastoma was strongly suspected, the significant smoking history notwithstanding. At surgery clear fluid from the cyst was aspirated. Although no clear nodule was identified cerebellar tissue in the area of the nodule was resected with the assistance of frameless neuronavigation. No discernable cyst wall was noted. Cytological examination of the fluid did not reveal any malignant cells. Histological examination of the putative mural nodule demonstrated only normal cerebellar architecture with neither malignant nor inflammatory cells. This amounted to a biopsy of the cyst wall revealing normal cerebellar architecture. It was believed that the absence of malignant cells in either the fluid or biopsy further supported the diagnosis of haemangioblastoma.

Somewhat expectedly (given the absence of pathological histology) the patient's symptoms recurred and the mural nodule and cyst were again demonstrated on imaging studies some months later. Given the inability to resect the mural nodule at the first surgery, and the patient's reluctance for repeat surgery, the patient was referred for stereotactic radio surgical ablation of the mural nodule. The MRI on this occasion did show some cyst enhancement but this was erroneously thought to be post-surgical in nature. The patient responded well to radiosurgery and a contrasted CT scan some three months afterward appeared to show diminution in both the sizes of the nodule and the cyst.

Approximately 10 months after the initial surgery the patient retuned with severe ataxia. MRI showed the cyst to be larger and more irregular with clear unequivocal enhancement. Even

on this occasion the possibility of an alternate diagnosis was not strongly entertained, the changed appearance believed to be radiation induced; consequently the patient was returned to the operating theatre with the intention of simple cyst drainage in the belief that radiation induced complete obliteration of the nodule would only become evident at a later stage.. However, at the second surgery hemorrhagic cystic fluid was encountered as well as a well-formed cyst wall which was excised in total. Histological examination on this occasion revealed a metastatic adenocarcinoma. Extensive subsequent systemic investigation, including a lung biopsy, failed to reveal a primary tumour. The patient refused chemotherapy and died 15 months (January 2019) after the initial surgery as a result of complications of a femoral fracture.

Discussion

Haemangioblastomas are cytologically benign ,vascular neoplasms of the central nervous system(Resche1993) ; that occur sporadically or 3 to 4 times more commonly as a component of Von Hippel Lindau syndrome(VHL).In the brain haemangioblastomas occur overwhelmingly in the posterior fossa and predominantly in the cerebellum (Sung1982) ; and is the most common primary (benign)intra-axial posterior fossa tumour in adults(Greenberg2001).

The MRI appearance of haemangioblastomas range from wholly solid to cystic with no discernable solid component/ nodule. The most common appearance is a non -enhancing cystic mass with an enhancing mural nodule of varying sizes (Goyal 2016, Kim 2020). It has been suggested that the appearance of a peripheral posterior fossa cyst with a mural nodule supplied by enlarged vessels on angiography may be pathognomonic for haemangioblastoma (Lee 1989, Kim 2020). The cyst wall generally does not enhance on either CT or MRI and enhancement should suggest an alternate diagnosis (Ho1992).

Surgical resection of sporadic haemangioblastomas, when possible, is curative. In the instance of cystic lesions resection of the mural nodule is sufficient and the cyst wall does not require excision.

Stereotactic radiosurgery (SRS) as a treatment option for haemangioblastoma has been previously described, (Patrice 1996, Kano 2015, Goyal 2016), and has been used for close to three decades now. In most instances, patients referred for SRS had had previous incomplete or failed resections or were confirmed to have VHL syndrome. Primary SRS, even for non-syndromic haemangioblastomas has nevertheless been reported (Kano 2008, 2015) the diagnosis being made in these instances by MRI and angiography. Although angiography was not done in the current case it was felt that the negative surgical findings supported the MRI diagnosis of haemangioblastoma.

Conclusion

With the benefit of hindsight, the correct diagnosis could have been reached had surgery been performed earlier instead of radiosurgery. The lesion was, in retrospect, an adenocarcinoma in evolution with missed radiological clues apparent before the correct diagnosis was eventually reached.

This case is a timely reminder that representative histological examination remains essential in the diagnosis, and subsequent treatment, of most brain tumours .Notable exceptions include advanced glioblastoma (White 1996) and syndromic haemangioblastomas .However "classical" the radiological appearance of a brain tumour , the assertion of Friedman *et al*(1989) that 'tissue diagnosis remains the foundation on which appropriate therapy can be based…..' remains as true today as it did more than 30 years ago.

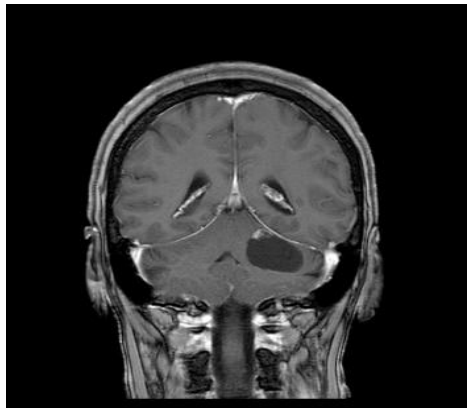

Figure 1 Initial Contrasted MRI

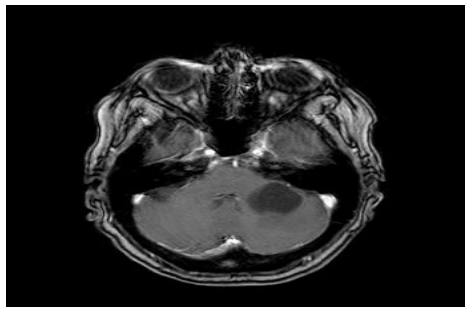

Figure 2 Initial axial contrasted MRI

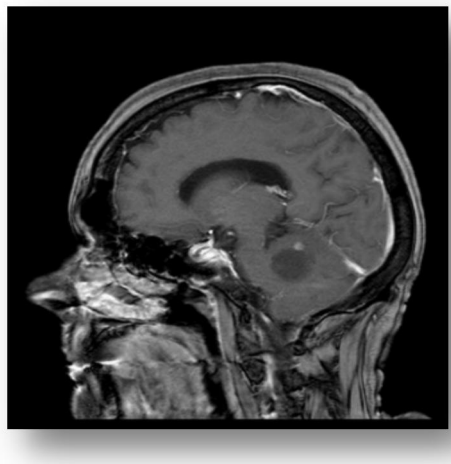

Figure 3 Initial contrasted sagittal MRI

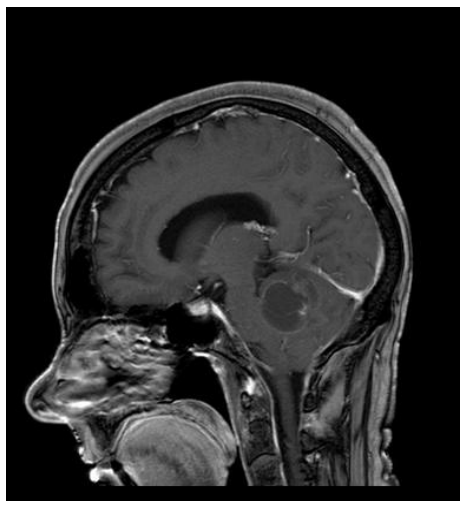

Figure 4 Contrasted MRI done prior to SRS

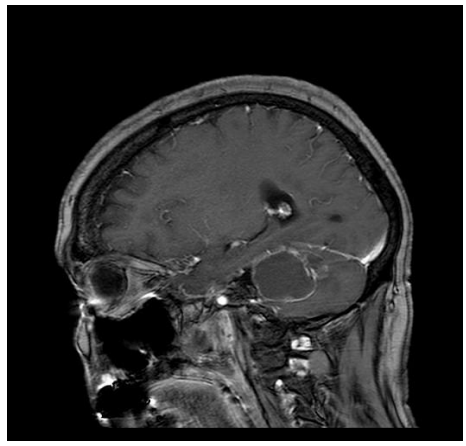

Figure 5 Contrasted MRI done prior to final surgery

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