CASE REPORT

Atlantoaxial synovial cyst causing isolated unilateral hypoglossal nerve paralysis

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ABSTRACT. Cervical synovial cysts are rare entities that are, most of the time, asymptomatic. They can cause nerve root or spinal cord compression, especially when acute haemorrhage or a marked increase in size occurs. Isolated unilateral hypoglossal nerve paralysis caused by compression of its cisternal segment is also an extremely rare condition. We report the case of a 51-year-old woman who presented with dysarthria and tongue fasciculation. MRI revealed an atlantoaxial synovial cyst that extended cranially through the hypoglossal canal and compressed the fibres of the left XII nerve on its cisternal segment. To our knowledge, this is the first case report of XII nerve paralysis being caused by an atlantoaxial synovial cyst.

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Spinal synovial cysts are usually found in the lower lumbar spine, being rare in the cervical spine [1–5]. Isolated unilateral hypoglossal nerve paralysis is also a very rare condition [6]. We present a case of an atlantoaxial synovial cyst causing isolated unilateral compressive hypoglossal nerve paralysis in a 51-year-old woman. To the best of our knowledge, this is the first report of a XII nerve paralysis caused by an atlantoaxial synovial cyst.

Case report

A 51-year-old woman presented with a one year history of dysarthria and tongue fasciculation, as well as chronic headaches. Physical examination revealed isolated left hypoglossal nerve palsy; no other abnormalities were seen in the neurological evaluation.

The patient underwent angiotomography of the cerebral vessels, which showed a discrete asymmetry of the hypoglossal canals (Figure 1), the left one being the larger. No other significant alterations were seen.

Contrast-enhanced brain MRI showed a lobulated cystic mass originating in the left atlantoaxial joint, which extended superiorly and insinuated through the ipsilateral hypoglossal canal. The mass had a signal intensity similar to the cerebrospinal fluid, being hypointense on T_1 weighted images and hyperintense on T_2 weighted images with no contrast enhancement. The three-dimensional constructive interference in steady state (CISS) sequence better depicted the sharp margins of the lesion and demonstrated more clearly the compression of the left hypoglossal nerve (Figure 2). In addition, the T_1 weighted images showed high signal

intensity on the left half of the tongue, representing fatty replacement as a consequence of atrophy.

The surgical treatment of the lesion was indicated after the diagnosis of a presumed atlantoaxial synovial cyst was suggested. However, the patient refused surgery and remains in follow-up for one year without progression of symptoms.

Discussion

Synovial cysts are cystic dilatations of the synovial sheaths that are found in joints and tendon sheaths [3], especially in hand and wrist joints [7]. The aetiology of spinal synovial cysts is unclear, and many theories have been proposed to explain their origin, such as trauma, metaplasia, the presence of developmental rests and excess stress at the facet joints with herniation of synovial tissue [3, 5, 8]. These cysts are assumed to be degenerative [1] and are considered an extrusion of the synovium through a capsular defect from a degenerated or unstable facet joint [5, 9]. In some cases, history of acute trauma is mentioned [10]. The cysts are very rare in the spine [3] and manifest during the fifth and sixth decades with no gender predominance [5]. Most spinal cord synovial cysts are asymptomatic, but they may cause nerve root or spinal cord compression, especially when acute haemorrhage or marked increase in size occurs [1, 5, 11]. Spinal synovial cysts are more frequent in the lower lumbar spine, being extremely rare in the cervical spine [1–5]. In this report, a 51-year-old woman with a presumed atlantoaxial synovial cyst had symptoms related to compression of the left hypoglossal nerve; no previous history of trauma or rheumatoid arthritis was referred.

Birch et al [1] reported a series of five patients with atlantoaxial synovial cysts and reviewed five additional

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Figure 1. Cerebral angiotomography. (a) The normal right hypoglossal canal is observed (arrowhead). (b) A discrete enlargement of the left hypoglossal canal (arrowhead).

cases from the literature. The patients, who ranged in age from 60 to 85 years, included three men and seven women with no history of rheumatoid arthritis or trauma. In all patients, the clinical presentation was related to cervical spinal cord compression (motor and/or sensory alterations), with an average duration of five months. None of the patients had signs or symptoms related to the hypoglossal or to any other cranial nerve.

In the majority of cases, synovial cysts can be accurately differentiated from other lesions by their neuroanatomical location and specific MRI signal characteristics and contrast enhancement pattern [1–4, 11, 12]. The classical appearance of a spinal synovial cyst is a well-defined, sharply margined lesion arising in a facet joint, presenting signal intensity similar to the cerebrospinal fluid (iso/hypointense on T_1 and hyperintense on T_2 weighted images) and no enhancement after intravenous gadolinium administration. In some cases, a slightly hyperintense signal in relation to the cerebrospinal fluid is seen owing to high protein content or haemorrhage [11]. Corroborating the imaging findings described above, our case showed a well-defined lobulated cystic mass in the left atlantoaxial joint, which was hypointense on T_1 and hyperintense on T_2 weighted images with no contrast enhancement. Considering the anatomical topography, the differential diagnosis of the lesion presented in this report should include hypoglossal nerve schwannomas, posterior fossa meningiomas and jugulotympanic paraganglioma. Because the MR images of these tumours differ from those seen in our case, we suggested the diagnosis of atlantoaxial synovial cyst for this 51-year-old woman.

The hypoglossal nerve is a pure motor nerve that supplies the intrinsic and extrinsic muscles of the tongue. The fibres of this nerve exit between the olive and the pyramid of the bulb and, with a short cisternal segment, pass in the direction of the hypoglossal canal in the occipital bone. This canal lies posteromedial and inferior

to the jugular foramen. Many diseases can cause unilateral tongue atrophy owing to central or, more commonly, peripheral involvement of the hypoglossal nerve. More commonly, the hypoglossal nerve is involved along with other cranial nerves, especially the glossopharyngeal, vagal and spinal accessory nerves. Isolated unilateral hypoglossal nerve paralysis caused by clear mechanical compression of the nerve is a very rare phenomenon [13, 14]. Mujic et al [6] reviewed the causes of unilateral compressive hypoglossal neuropathy and suggested the following differential diagnosis: dural arteriovenous fistula, enlarged emissary vein in the hypoglossal canal, aneurysm of the stump of persistent hypoglossal artery, fracture of occipital condyle and an arachnoid cyst. To the best of our knowledge, there is only one case [6] of spinal synovial cyst compressing the hypoglossal nerve, and no previous reports of atlantoaxial synovial cyst causing isolated hypoglossal nerve paralysis.

In our case, an extensive atlantoaxial synovial cyst projected cranially through the left hypoglossal canal into the pontocerebellar angle, compressing the fibres of the XII nerve on its cisternal segment. As the lesion showed all the classical features of a cystic-appearing mass and originated in the atlantoaxial joint, the diagnosis of atlantoaxial synovial cyst was suggested. The fact that there were no changes in the lesion during the follow-up examinations also favoured this diagnosis. As the patient presented with a one year history of dysarthria and tongue fasciculation, we could also observe a muscular atrophy of the tongue with fatty replacement of the left half.

In conclusion, the association of atlantoaxial synovial cyst and isolated unilateral compressive hypoglossal nerve paralysis is rarely seen. In this report we presented the second case of twelfth nerve paralysis caused by a presumed synovial cyst and the first caused by an

Figure 2. (a, c) Axial CISS-3D MRI with coronal reconstruction (b) showing an expansive lesion with sharp margins at the left atlantoaxial joint (arrow in a). This lesion extends superiorly (arrowheads) and insinuates into the hypoglossal canal (arrow in c).

atlantoaxial synovial cyst. We recommend that cervical synovial cysts should be included in the differential diagnosis of isolated unilateral hypoglossal nerve palsy.

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