

Anterior, extracanal, cervical spine osteochondroma associated with DISH: description of a very rare tumor causing bilateral vocal cord paralysis, laryngeal compression and dysphagia. Case report and review of the literature

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Abstract. – INTRODUCTION: Osteochondromas are common benign bone tumors, rarely involving the spine.

BACKGROUND: Osteochondroma account for about 35% of bone benign tumors and 9% of all bone tumors. Spinal involvement is described in 1%-4% of cases and their origin from the anterior surface of cervical vertebral bodies is exceedingly rare.

AIM: We describe the rare case of an osteochondroma arising from the anterior surface of the C4 and C5 vertebral bodies, and not involving the spinal canal, in a 68-year-old male patient suffering from Diffuse Idiopathic Skeletal Hyperostosis (DISH).

MATERIALS AND METHODS: The patient presented with acute onset of respiratory distress due to laryngeal compression exerted by the lesion, dysphagia and paralysis of left vocal cord. Imaging revealed the unusual lesion compressing and dislocating the air ways. An anterior approach to cervical spine was performed to remove the tumor.

RESULTS: Postoperatively, the patient sustained a progressive improvement of respiratory function and recovery of the vocal cord paralysis.

DISCUSSION: We believe that this case holds some interesting peculiarities: firstly, the anterior location of a cervical osteochondroma could be considered exceptional; yet, the resulting clinical picture should be remembered for differential diagnosis. Secondly, we hypothesized a possible correlation between the tumorigenesis of osteochondroma and the co-existence of DISH. Indeed, the abnormal bone turnover in cervical segments due to DISH could explain the unusual occurrence of osteochondromas in adult age.

CONCLUSIONS: The occurrence of an osteochondroma should be considered in patients suffering from DISH and harbouring large osteophytes.

Key Words:

Spinal osteochondroma, Cervical spine, DISH, Laryngeal compression, Dysphagia.

Introduction

Osteochondroma is a common benign bone tumor accounting for 35% of all bone tumors¹, usually involving long bones. The spinal involvement is extremely rare, representing 1 to 4% of overall cases. The first papers describing a cervical spine osteochondroma have been reported over forty years²⁻⁴. Subsequently, several authors reported cases of cervical osteochondromas, highlighting a large spectrum of different clinical pictures⁵. According to the site of the lesion, the most common clinical presentation is related to the involvement of nervous structures, with myelopathy, radiculopathy and, occasionally, also neck pain⁶. Such clinical picture is secondary to the frequent localization of the tumor in the vertebral body posterior elements⁷. Vertebral artery occlusion⁸, palpable neck mass⁹, dysphagia and sleep apnea¹⁰ as well as occipital nerve neuralgia¹¹ have been reported as uncommon presentations of osteochondromas involving cervical spine.

Usually, osteochondromas arising from the spine are multiple, in particular if associated with hereditary multiple exostosis. Conversely, the solitary osteochondroma of the spine is a less frequent entity. The association between the occurrence of osteochondromas and hereditary multiple exostosis has been already described, especially in young patients^{12,13}. Even in these

latter cases, the clinical presentation is usually characterized by spinal cord or nerve root compression. Conversely, the occurrence of the osteochondroma in elderly patients still remains unclear and the association with possible predisposing factors has not been investigated.

Aim

The aim of this report is to describe a case of an anteriorly sited cervical spine osteochondroma in a 68-years old man affected by diffuse idiopathic skeletal hyperostosis (DISH), determining a severe respiratory distress due to laryngeal compression and unilateral vocal cord dysfunction. The anterior location of cervical osteochondromas is certainly exceptional: a literature review revealed only three cases of anteriorly located osteochondroma of the cervical spine^{10,14,15}. However previous cases presented differences in their management, and in one case no histological examination was performed.

Moreover, to our best knowledge, the association of cervical osteochondroma and DISH in elderly age has not been discussed in the literature.

Case Report

History and Clinical Examination

A 68-year old man presented to the Emergency Department with acute onset of dyspnea and respiratory distress; dysphagia was reported to have been present since few weeks. His symptoms were diagnosed to be secondary to a rapidly progressing airway obstruction, which was urgently treated by tracheostomy. A computed tomography (CT) scan of the neck and a magnetic resonance imaging (MRI) of the cervical spine revealed an extrinsic laryngeal compression, with left-side dislocation of the laryngo-tracheal axis, due to the presence of a “solid” mass arising from the anterior surface of C4 and C5 vertebral bodies (Figures 1, 2 A-B, 3 A). This condition coexisted with a diffuse ossification of the anterior longitudinal ligament, extending to the lower thoracic segments. The patient also underwent an endoscopic examination of the larynx, which revealed a significant reduction of respiratory space and a bilateral paralysis of the vocal cords (Figure 5 A). Upon presentation no neurological abnormalities were found.

Surgical procedure

The patient underwent surgery in order to remove the cervical mass, decompress the airways and restore normal respiratory function. A standard antero-lateral approach to the cervical spine was performed to reach the anterior surface of the spinal column. The pre-vertebral longus colli muscles appeared laterally displaced by the lesion. After dissecting these muscles off the lesion, a hard bone-like mass, mushroom-shaped, was exposed. Using either blunt dissection and scalpels, the lesion was separated from the surface of C4 and C5 vertebral bodies. The resection was deliberately limited to the mass causing compression on air ways in order to limit complications (Figure 4 A). After removing the tumor, the abnormal ossification of the anterior longitudinal ligament and the hyperostosis at adjacent vertebral segments were clearly visualized (Figures 2 A-B, 3 A). In view of his neurological examination (i.e. absence of overt abnormalities), it was decided not to perform any microdissectomy and fusion.

Outcome

Histological examination, integrated by immunochemistry analysis, confirmed that the mass was an osteochondroma (Figure 4 B-C).

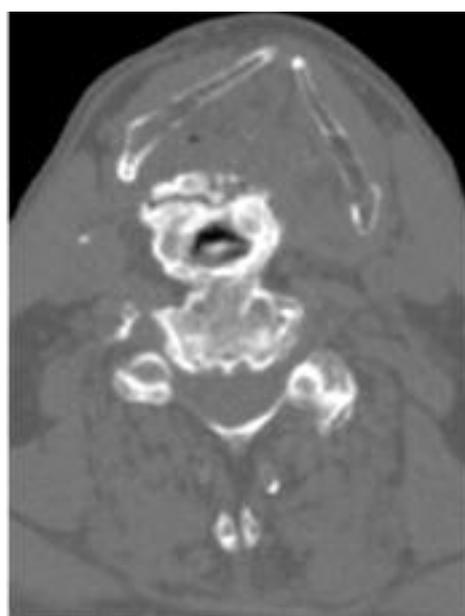


Figure 1. Preoperative, axial CT image shows a large bone solid mass arising from anterior surface of C4 and dislocating laterally and anteriorly the esophagus and the trachea.

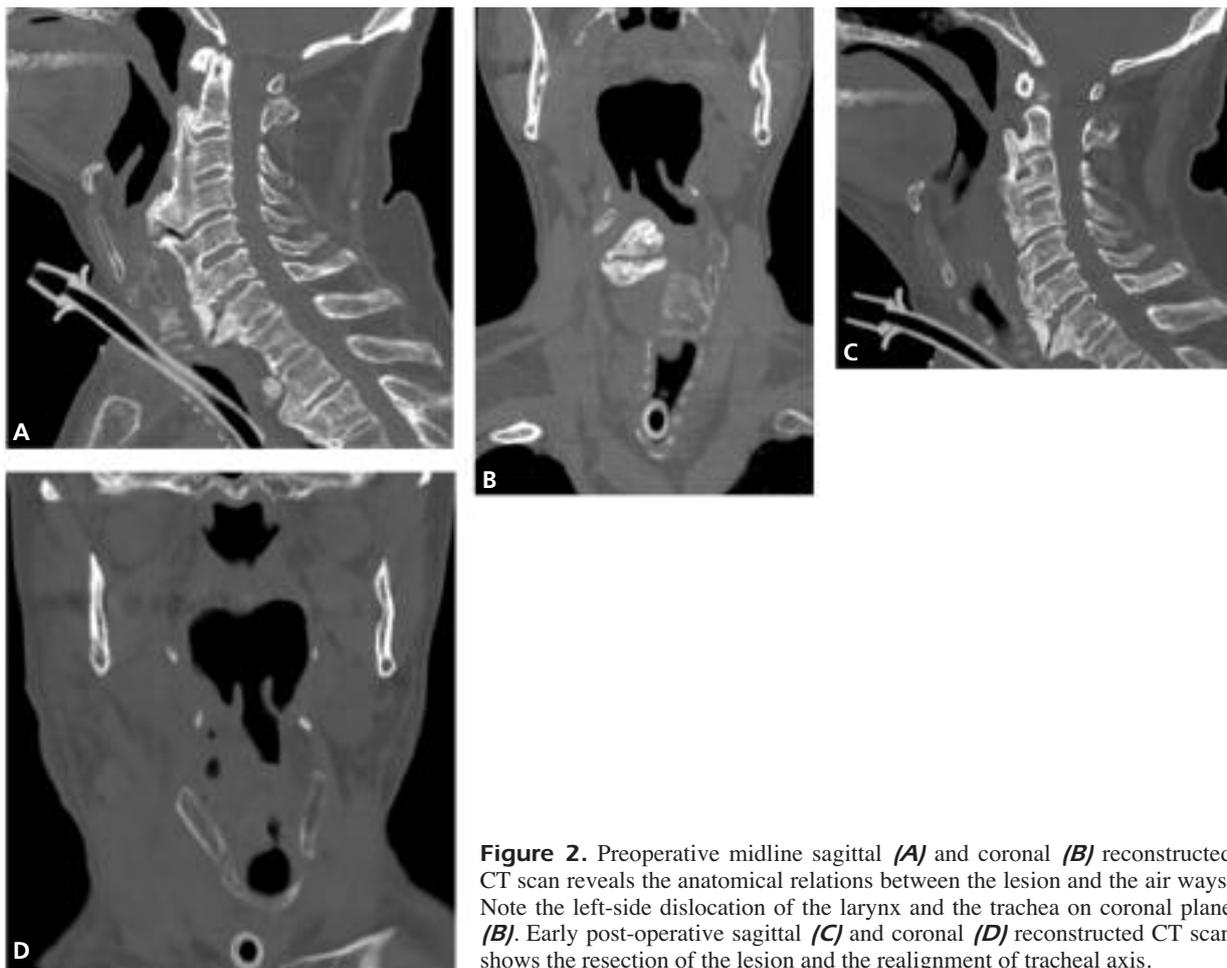


Figure 2. Preoperative midline sagittal (*A*) and coronal (*B*) reconstructed CT scan reveals the anatomical relations between the lesion and the air ways. Note the left-side dislocation of the larynx and the trachea on coronal plane (*B*). Early post-operative sagittal (*C*) and coronal (*D*) reconstructed CT scan shows the resection of the lesion and the realignment of tracheal axis.

The patient did not experience any post-operative complication. Post-operative CT scan (Figure 2 C-D) showed the removal of the lesion and a good decompression of the airways and esophagus. A slight dysphagia persisted, but this did not stop the patient from restarting oral semi-liquid intake. A post-operative endoscopic examination of the larynx was performed, one week after surgery. Interestingly, a good recovery of the paralysis of the left vocal cord was documented, together with initial recovery of the paralysis of the right one and restoration of normal respiratory space. Consequently the tracheostomy was removed without consequences 30 days post-operatively. A 3 months follow-up MRI showed no recurrences and confirmed the tumor resection (Figure 3 B). Moreover, laryngoscopic follow-up examinations documented a progressive restoration of normal vocal chords motility within 8 months (Figure 5 B).

Discussion

Osteochondromas are benign bone tumors usually involving long bones, with onset in childhood and adolescence, representing about 8.5% of all bone tumors and about 36% of benign ones^{1,16}. Their incidence seems to decrease with age, even if a late onset in adults and elderly has been described¹⁷⁻¹⁹. In 2010, Gunay et al²⁰ reported a small series of six patients suffering from spinal osteochondroma. They described the clinical and radiological features of this pathology, highlighting the importance of correct pre-operative management and surgical treatment. Indeed, because of their rarity, osteochondromas are often scotomized among differential diagnostic options when a spinal lesion is radiologically demonstrated. Despite being small, Gunay's series is one of the largest in the literature together with the series reported by Zaijun et al²¹ and by Lotfinia et al⁷. In Zaijun's clinical series, 14 pa-

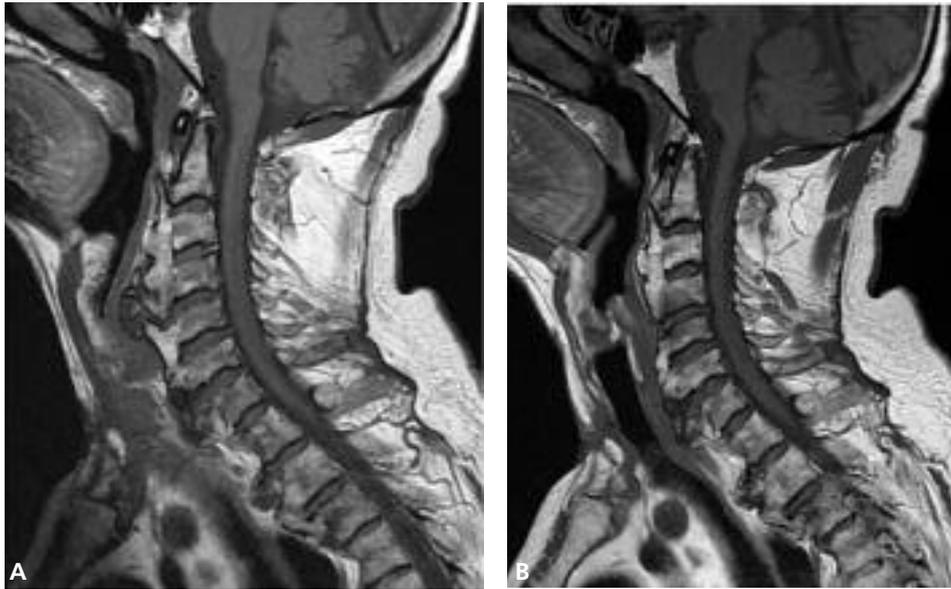


Figure 3. Preoperative sagittal, T1-weighted MRI image **(A)** shows the mass arising from the anterior surface of C4 and C5 vertebral bodies, excluding spinal intracanal involvement. A diffuse ossification of anterior longitudinal ligament is visible. 3-months follow-up sagittal, T1-weighted MRI image **(B)** confirmed the removal of the lesion and the restoration of respiratory space.

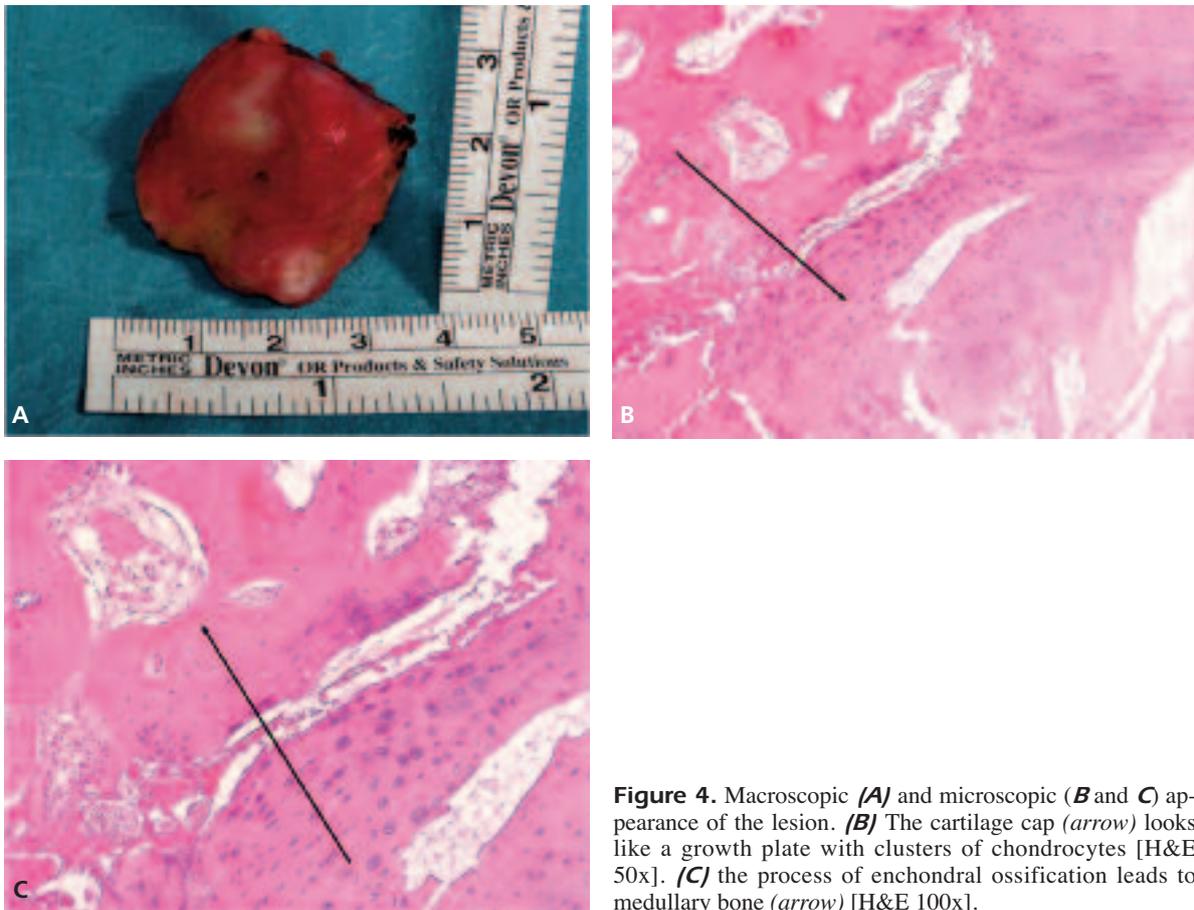


Figure 4. Macroscopic **(A)** and microscopic **(B and C)** appearance of the lesion. **(B)** The cartilage cap (*arrow*) looks like a growth plate with clusters of chondrocytes [H&E 50x]. **(C)** the process of enchondral ossification leads to medullary bone (*arrow*) [H&E 100x].

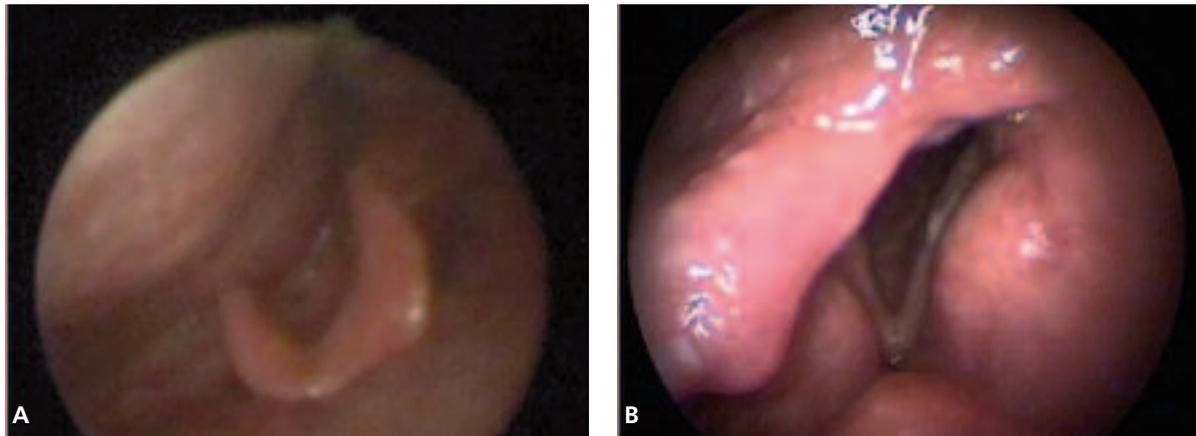


Figure 5. Preoperative laryngoscopy (**A**) reveals the extrinsic compression on the hypopharynx with severe reduction of glottis space; the vocal chords are not visible. 8-months follow up laryngoscopy (**B**) shows the complete restoration of respiratory space and the recovery of the vocal chords function.

tients affected by spinal osteochondroma were included; 8 of these patients presented a localization of the tumor in the cervical spine. Interestingly, the authors described the co-existence of a condition of hereditary multiple exostoses in two patients and a malignant sarcomatous transformation in two additional cases of recurrence. Similarly, Lotfinia et al⁷ reported a series of 8 patients surgically treated for a spinal osteochondroma (3 involving cervical segments). A correlation with multiple hereditary exostoses was found in 33% of cases equally distributed in cervical, thoracic and lumbar spine. Previously, in 2005, Gille et al²², reported another small series composed by 6 patients surgically treated and followed for six years. They reviewed also literature concerning spinal osteochondromas, finding overall 150 cases of solitary osteochondroma; six of those (4%) presenting local recurrence and four (2.7%) malignant transformation. The focus on previously published papers on spinal osteochondromas reveals some interesting points. Provided that spinal osteochondromas are rare lesions, it seems important the correlation with congenital conditions like hereditary multiple exostoses. It is also relevant, with regard to management and therapies, the impact of the possible malignant transformation of this pathology. Indeed, as recommended by Zaijun²¹, gross total resection has to be attempted in order to prevent the risk of recurrence. Besides the above few small series, only isolated cases of spinal osteochondromas have been reported in the literature.

We believe that the present case presents peculiar features, which make it different from the others

previously published. The first aspect concerns the location of the tumor. Atypical location of spinal osteochondromas in the cervical segment has already been described. Schomacher et al²³ reported a case of osteochondroma arising from the right atlanto-axial joint (C1-C2), with a medial and posterior development, causing spinal cord compression. A secondary involvement of the high cervical spine was described by Lopez-Flores et al²⁴: they reported on a 35-year old male suffering from an osteochondroma arising from the right occipital lateral condyle. This tumor presented a longitudinal extension both upwards and downwards, involving, respectively, the foramen magnum and C1-C2 joint. However, this case has to be considered among skull base osteochondromas, because of its origin from the condyle. Osteochondromas can also originate from the spinous processes of cervical spine, either in pediatric patients, as described by Chon et al²⁵, or in a young adults, as reported by Bonic et al⁹. Our case is characterized for an atypical development of the tumor, which originated from the anterior surface of vertebral bodies at C4 and C5 levels. The clinical picture and the onset of symptoms were consequential to its location; indeed our patient did not present any motor or sensitive deficits because spinal cord and nerve roots were not involved. The peculiar presentation was due to the compression exerted by the mass on the upper respiratory tract and to the resulting strain on the left recurrent laryngeal nerve.

An origin of osteochondromas from the anterior surface of the cervical spine has been previously reported in three papers. In 2005, Grivas et al¹⁴ described a patient presenting with dysphagia

and neck pain due to a bony mass, histologically defined as osteochondroma, located on the antero-lateral surface of C7 vertebral body. They stressed the importance of the surgical management of this condition, in order to definitively solve the symptoms. Wang and Chou, in 2009¹⁰, reported the case of a sporadic osteochondroma arising from the anterior arch of the C1 vertebra causing dysphagia and obstructive apnea in a pediatric patient. Despite the anterior location, this case cannot be considered similar to ours, because of the peculiarities of the first two cervical vertebrae which are anatomically and physiologically different from the vertebrae of subaxial cervical spine. Finally, Reckelhoff et al¹⁵ recently reported an osteochondroma arising from the anterior surface of C4 vertebral body, presenting with neck pain and treated by chiropractic treatments. However, the latter authors did not clarify if a histological confirmation of the lesion had been performed. We want to emphasize that histological examination still remains the only and definitive tool for a correct diagnosis of osteochondromas, despite advanced imaging offers valuable and useful information. Under such circumstance, our report is only the second describing a cervical spine osteochondroma arising from the anterior vertebral body's surface, and the first describing a "pure" anterior origin of the tumor.

The second important feature to highlight in our case is the correlation between tumor development and presence of diffuse idiopathic skeletal hyperostosis (DISH). DISH, also called with the eponym Forestier's disease, is a clinical syndrome characterized by the abnormal formation of osteophytes involving the spine, with ossification of the paraspinal muscles and ligaments. This condition was originally described by Forestier and Rotes-Querol in 1950²⁶ as a calcification of anterior longitudinal ligaments. Subsequently in 1975, Resnick²⁷ defined the acronym of "DISH" to describe the syndrome. He together with Niwayama²⁸ encoded also, two years later, diagnostic criteria for Forestier's disease. These included:

1. Longitudinal anterior ligament calcification involving at least four contiguous vertebrae;
2. Absence of degenerative changes of the affected disks;
3. Absence of ankylosis of the joints and sacroiliac arthrosis phenomena.

DISH is a common condition in the aging spine, often associated with large anterior osteophytes of the cervical spine, which may cause symptoms²⁹. Moreover, some authors described Forestier's dis-

ease as cause of dysphagia or laryngeal compression, discussing medical and surgical management^{30,31}. The mechanisms of onset of dysphagia in DISH is not only related to mechanical compression, but also include esophageal and/or pharyngeal inflammation with edema and fibrosis and cricopharyngeal spasm also due to chronic irritation³². Surgical treatment is advocated only after failure of conservative therapies. Conversely, in absence of relevant clinical signs due to the presence of DISH surgery is not considered mandatory.

The patient we describe presented all three inclusion criteria for diagnosis of DISH and pre-operative radiological assessment did not suggest a diagnosis of neoplasm but rather suggested a classical picture of a large osteophyte in the context of DISH. Indeed, we believe that the appearance of an osteochondroma in a patient affected by DISH could be related. We hypothesize that the alteration of bone turnover causing DISH could be a trigger mechanism for uncontrolled cell proliferation. This may lead to the formation and growth of the tumor also after the completion of physiological bone growth. Yagi et al²¹ have made a similar hypothesis advocating psoriatic arthritis as possible mechanism triggering the enlargement of osteochondromas in elderly patients. Therefore, our case suggests that surgical treatment could also play an important diagnostic role in presence of symptomatic DISH, particularly if we consider the possible progression to malignancy of osteochondromas.

Conclusions

In patients suffering from DISH and harbouring large osteophytes, the possible development of an osteochondroma should be considered. In these patients, if symptomatic, surgical treatment is recommended in order to reduce the mass effects and to confirm the diagnosis. A pathophysiological correlation between the late onset of osteochondroma and the co-existence of DISH could be hypothesized.

Disclosure

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Conflict of Interest

The Authors declare that there are no conflicts of interest.

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