La Prensa Medica Argentina

Case Report

A Rare Case of Right Paravertebral Myoepithelioma with Intradural Extension Causing Cord Compression

Sachin Kale[1], Manit Arora[1*], Sameer Mansukhani[1], Rajendraprasad R. Butala[1], Prasad Chaudhari[1] and Sanjay Dhar[1]

[1]Department of Orthopaedics, Padmashree Dr. DY Patil Hospital and Research Centre, Navi Mumbai, India

Corresponding author: Dr. Manit Arora MBBS (Hons), MS(Ortho), Department of Orthopaedics, Padmashree Dr. DY Patil Hospital and Research Centre, Navi Mumbai, India, Tel: +918452846005; E-mail: manit_arora@hotmail.com

Rec date: Mar 27, 2014 **Acc Date:** May 26, 2014 **Pub date:** May 28, 2014

Abstract

Myoepithelial tumors occur mainly in the salivary glands, the sweat glands or the breast. There has been no case of paravertebral myoepithelioma with intradural extension reported in the literature. We present a case of a large right paravertebral myoepithelioma with intradural extension causing cord compression that was referred to our institution for palliative cord decompression.

Introduction

Myopethilelial tumors are most frequently encountered in the salivary glands, sweat glands and mammary glands, in descending order of frequency [1,2]. These neoplasms usually show morphological characteristics resembeling those of myoepithelial cells in the affected gland system. Little is understood about these rare neoplasms, possibly due to their paucity of occurrence and reporting. To the authors' knowledge, there are no previously reported cases of intradural spinal involvement of myoepithelioma. We present a case of a large right paravertebral myoepithelioma with intradural extension causing cord compression that was referred to our institution for palliative cord decompression.

Case Report

A 49 year old male shopkeeper presented to our institution for palliative decompression surgery of the spine. The patient had a three year history of back pain with radiation into bilateral lower limbs associated with right sided radiculopathy and perianal parasthesia, and constitutional symptoms (weight loss, anorexia and lethargy).

The patient gave a three year history of lower dorsal and upper lumbar back pain of dull aching nature present at rest and unrelieved by analgesia. The patient described bilateral radiation of the pain into the lower limbs associated with bilateral parasthesias and weakness. There was a positive history of perianal parasthesia and difficulty urinating. The pain and parasthesia had been worsening over the last 6 months. The patient also had constitutional symptoms of weight loss (5 kg over 3 years), anorexia and lethargy.

The patient gave a negative history of bowel or bladder incontinence, fevers, night sweats, trauma or falls. There was no contact with TB or HIV carriers in the last five years. There was a positive history of unprotected sexual intercourse with the spouse.

On examination, the patient had a large globular swelling in the right paraspinal region, and right upper and lower abdominal quadrants with an overlying scar from previous failed attempted excision (Figure 1). There was severe and painful restriction of spinal movements with a finger-floor distance of greater than 20 cm and a modified Schouber's test of 2cm. Power in the left lower limb was of grade 5, and in the right lower limb of grade 4 proximally and distally. The patient had loss of sensation below the level of L1 bilaterally including the perianal region. Anal wink and bulbocavernous reflexes were preserved.

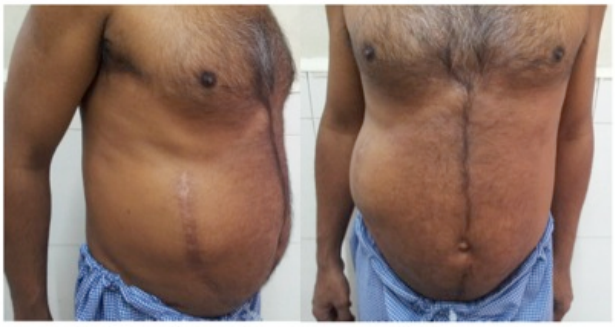

Figure 1: Clinical photographs of the patient.

The patient had previously (3 years ago) been diagnosed with right paravertebral myoepithelioma extending from D11 to L2 vertebra at a specialist oncology centre. Excision was attempted initially with 2 attempts at radiofrequency ablation. Histopathology and immunochemistry techniques at the time confirmed myoepithelioma. (multinodularmyxoid tumor with epithelia cells; no physaliphorous cells; no significant mitoses/necrosis; Vimentin and p63 marker positive; S100, CK 5/6 and brachial CK markers were negative). However, tumor had recurred within 6 months with satellite nodules in the right psoas muscle and intradural extension causing cord compression. There was no evidence of metastases. The patient was referred to us for palliative decompression of the cord.

The patient's blood tests were unremarkable other than poor renal function (Sr. uric acid – 6.6mg/dl; Sr. globulin – 4.2mg/dl; Sr. creatinine - 1.9mg/dl; blood urea nitrogen – 30mg/dl). There was no leukocytosis, no anaemia of chronic disease and no evidence of marrow suppression or liver dysfunction.

Radiograph of the dorso-lumbar spine (Figure 2) and MR imaging (Figure 3) was consistent with a high grade sarcoma in the right paravertebral region extending from D10 to L5 vertebral level, with contiguous involvement and wedging of the L1 vertebra with intraspinal extension at the D12/L1 and L1/L2 levels causing compression of the conus and cauda equine roots.

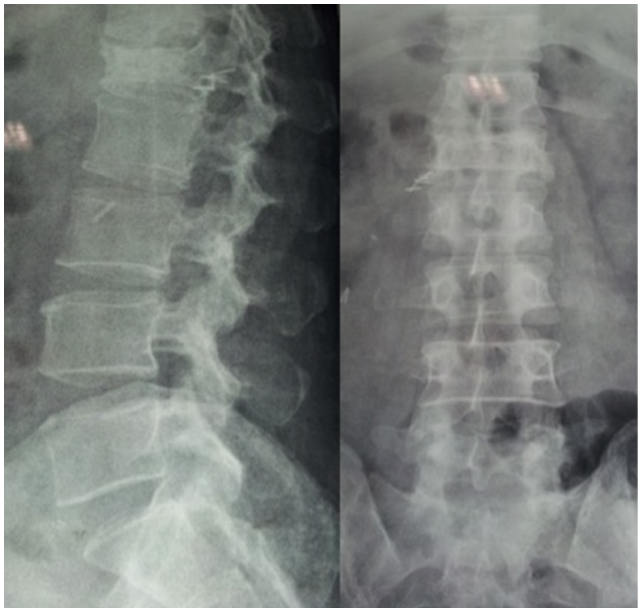

Figure 2: Anteroposterior and lateral radiographs of the dorso-lumbar spine showing destruction and wedging of the L1 vertebra with reduced disc space at D12/L1 and L1/2 with associated kyphosis.

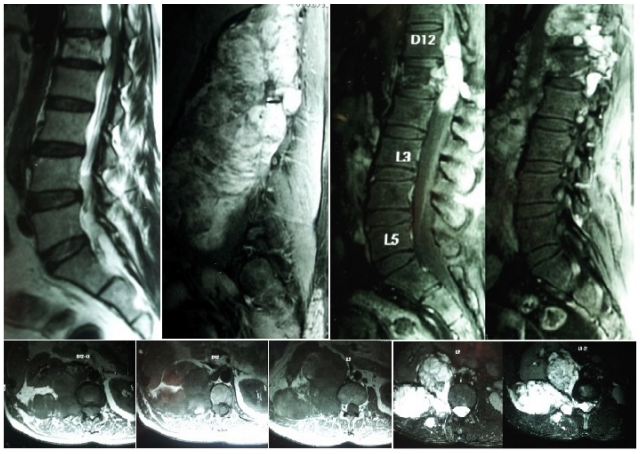

Figure 3: T1 and T2 weighted axial and saggital images showing a large multi-lobular right paravertebral mass extending from D10 to L5 vertebral level, with contiguous involvement and wedging of the L1 vertebra with intraspinal extension at the D12/L1 and L1/L2 levels causing compression of the conus and cauda equine roots.

We performed a decompressive laminectomy from D12 to L2 without any posterior instrumentation (Figure 4). The patient was managed post-operatively with rehabilitation using a Knight-Taylor brace when non-recumbent. No steroids were given to the patients in the pre or post-operative period. The patient had total recovery of motor and sensory function within 1 week and at 2 weeks follow-up was neurologically stable with near total recovery except for right L5 weakness of grade 4. The patient will be managed in a Knight-Taylor brace for an additional 4 weeks with gradual range of motion exercises for the spine and strengthening of the core musculature.

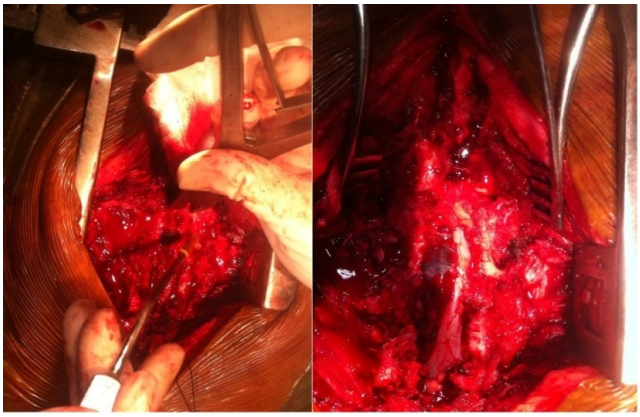

Figure 4: Intra-operative photographs of decompressive laminectomy.

Discussion

Myoepithelial cells are ectodermally derived contractile cells, routinely identified in many normal tissues with a secretory function such as major and minor salivary glands, lacrimal glands, sweat glands, breast and prostate [1,2]. Neoplasms composed primarily of myoepithelial cells are uncommon, accounting for less than 1% of all salivary gland tumors [3]. Most of these tumors are located in the parotid gland, with close to 100 cases reported in the literature for this site [4]. Other sites of involvement are less frequently reported. There is no report to our knowledge of a paravertebral myoepithelioma with intradural extension.

The term myoepithelioma was introduced by Sheldon in 1943. However, the precise criteria for inclusion of a tumor in this category remain controversial [3]. There is a wide variation in morphological and immunopheotypic characteristics amongst myoepitheliomas. Dardick et al. have proposed histopathologic guideline for myoepitheliomas [5].

Vimentin and S100 proteins are not usually present in normal myoepithelial cells and are very sensitive, but non-specific, markers of neoplastic myoepithelium [3]. The prognosis of benign myopethiloma would appear good, provided surgical excision is complete. Radiation therapy is used only when surgery is not considered feasible [3].

Conclusion

We present the first case report in the literature of a right paravertebral myoepithelioma with intradural extension, a rare neoplasm. The patient presented to us for surgical decompression secondary to intradural extension and cord compression of the tumor.

References

1. Barnes L, Appel BN, Perez H, El-Attar AM (1985) Myoepithelioma of the head and neck: case report and review. J Surg Oncol 28: 21-28.

2. Hamperl H (1970) The myothelia (myoepithelial cells). Normal state; regressive changes; hyperplasia; tumors. Curr Top Pathol 53: 161-220.
3. Ferri E, Pavon I, Armato E, Cavaleri S, Capuzzo P, et al. (2006) Myoepithelioma of a minor salivary gland of the cheek: case report. Acta Otorhinolaryngol Ital 26: 43-46.
4. Politi M, Toro C, Zerman N, Mariuzzi L, Robiony M (2005) Myoepithelioma of the parotid gland: Case report and review of literature. Oral Oncol Extra. 41:104–108.
5. Dardick I, Thomas MJ, van Nostrand AW (1989) Myoepithelioma--new concepts of histology and classification: a light and electron microscopic study. Ultrastruct Pathol 13: 187-224.