DOI: 10.7860/JCDR/2015/10644.5498

Case Report

Oncology Section

# Extradural Spinal Metastasis of Adenoid Cystic Carcinoma (ACC): A Case Report

VINOD KUMAR[1], RAJESH NAIR[2], SUNIL UPADHYAYA[3], BHAVNA NAYAL[4], ARJUN SHETTY[5]

## ABSTRACT

Adenoid cystic carcinoma (ACC) is a rare malignant tumour of the major salivary glands. It accounts for 10-15% of all salivary gland tumours and 1% of all head and neck tumours. Surgical resection followed by radiation is the choice of treatment for ACC. However, late loco-regional recurrence and metastasis is often seen emphasizing the importance of long-term follow-up.

We report an unusual case of extradural metastasis of ACC in the dorsal spine. The primary submandibular gland tumour was resected 11 y back. A recurrence had been detected two years prior to the occurrence of spinal metastasis. Surgical decompression was done which was followed by palliative radiotherapy. Patient is symptomatically better, ambulant and on regular follow-up.

**Keywords:** Adenoid cystic carcinoma, Compressive syndrome, Extradural spinal metastasis, Perineural invasion

## CASE REPORT

### History

A 45-year-old male presented to the neurosurgery out patient department with diffuse low back pain radiating to the right girdle since two months and inability to walk since two days. The pain was continuous in nature, exaggerated on activity and was persistent during the night with relief on taking analgesics. He had no associated co-morbid illness and or habituations. There was no history of trauma or tuberculosis.

Patient gave a past history of primary submandibular gland tumour resection 11 years back followed by radiation therapy. The diagnosis of ACC was rendered on the excised specimen (as per outside histopathology reports). He was advised an FDG-PET (Positron emission testing) scan, however differed the same. Nine years later, he developed a recurrent nodule at the previous scar and was detected to have recurrent disease on fine needle aspiration cytology. He underwent a wide local excision of the lesion with modified radical neck dissection (done at KMC, Manipal). The final histopathology was reported as ACC with negative margins and nodal metastasis (6 out of 12 isolated were positive for metastasis). He deferred palliative radiation therapy and was kept on regular follow-up.

### Examination

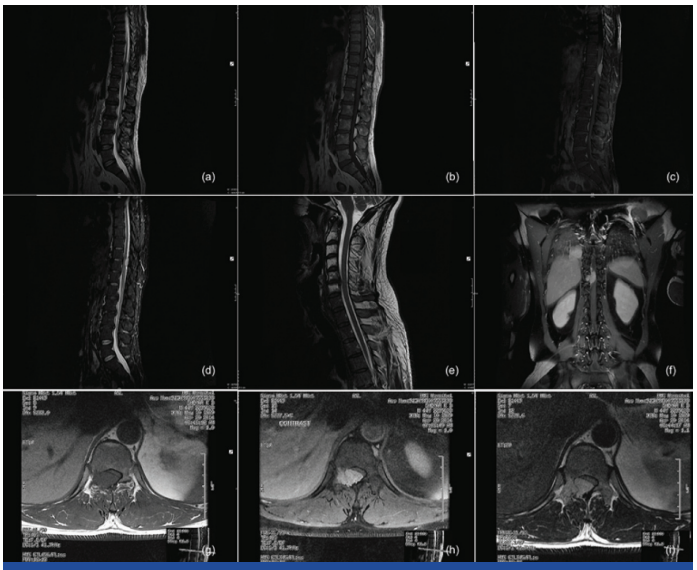
On clinical examination patient had mild spastic gait (Ashworth grade 4) [1] with bilateral lower limb spasticity. Power was 5/5 in both lower limbs. There was sensory loss below L1 level on the right side, and below L3 on the left side with gross involvement of the posterior column with no involvement of bowel or bladder. Bilateral knee and ankle jerk were brisk (3+) [1]. Mouth opening was restricted with Grade II trismus, intraoral examination revealing a densely fibrosed buccal mucosa [2] on the irradiated side. Rest of the oral cavity examination was unremarkable. He had a keloid formation over the operative site. On the outset, with the history of malignancy and an acute progression of lower limb motor function loss, our primary clinical diagnosis was (a) spinal metastasis, second was (b) epidural haematoma with compression over the traversing roots and less likely (c) bleed into a pre-existing meningioma.

### Investigation

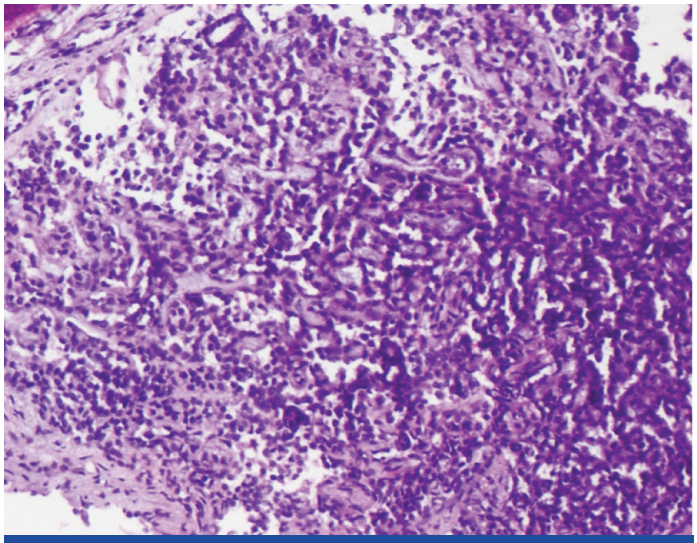
Magnetic resonance imaging (MRI) of the dorso-lumbar spine showed an extradural lesion at the level of D11 vertebra, hypointense on T1, hyperintense on T2 with contrast enhancement. The lesion measured 2.4 x 1.7 x1.5 cm with involvement of the right pedicle, articular pillar and lamina. The cord appeared displaced to the left with subtle T2w hyperintensity suggesting cord edema. Another enhancing T1 isointense and T2w hyperintense nodule measuring 3 x 2.7cm was seen in the postero-basal aspect of right lung suggestive of metastasis. Fatty marrow changes from C2 to C6 vertebral bodies were also noted indicating radiotherapy related changes [Table/Fig-1].

### Operative and Postoperative Course

The patient underwent D10-D11 laminectomy with surgical excision of the lesion. Histopathological diagnosis of metastatic adenoid cystic carcinoma was prefered [Table/Fig-2]. There was immediate




**[Table/Fig-1]:** Magnetic resonance image of the dorsolumbar spine – saggital (a-d) and axial sections (g-i) showing a lesion, hypointense on T1, hyperintense on T2 with contrast enhancement at the level of D11 vertebra. Fatty marrow changes from C2 to C6 vertebral bodies (saggital cuts) (e) indicating post radiation changes



**[Table/Fig-2]:** Photomicrograph showing bony trabeculae enclosing tumour cells in cribriform and adenoid pattern with luminal mucoid material (H&E, X200)

and significant symptomatic relief following the surgery. The patient was ambulant on the first postoperative day and was discharged on the 8th postoperative day. Palliative systemic chemotherapy and spinal radiation therapy was given and the patient is neurologically better on follow-up after eight months of spinal surgery.

## DISCUSSION

ACC is a rare tumour, which is difficult to treat. It occurs in the major and minor salivary glands of the skin and upper aero-digestive tract. Loco-regional recurrence and metastatic spread is well known. The common sites of metastasis are lungs, bone, liver and brain. Metastasis to the spinal vertebrae is extremely rare though perineural invasion in margin positive surgical resections have been noted in literature [3-6]. Typically, low back or neck pain is the commonest presentation of spinal metatstasis, followed by weakness, sensory loss, and bladder dysfunction [7].

ACC in the head and neck region is treated with surgical resection along with preoperative or postoperative irradiation. This reduces loco-regional recurrence and increases disease free survival rates [8,9]. Metastasis to the lymph nodes is rare and hence neck dissection is usually not warranted [10].

Histologically ACC has three different variants: glandular (cribriform), tubular and solid. Mitotic figures are generally scarce in cribriform and tubular; however these are seen frequently in the solid variant and have the worst prognosis [11]. There seems to be a direct co-relation between the histological pattern and prognosis in ACC [11,12].

The route of tumour spread from submandibular salivary gland to the spinal vertebrae remains debatable. The proposed routes are haematogenous metastases (via the venous drainage into the batson's venous plexus and then into the arterial arcade) or perineural invasion. The latter correlates with craniocaudal progression of the disease. Haematogenous metastasis to the brain is rare, though they have been documented in literature [5].

The median time to locoregional recurrence is two years and distant metastasis is three years with a three and five year disease free survival rates being 64.3% and 36.1% respectively [6].

High incidence of loco-regional recurrence and distant metastasis of ACC mandates prompt diagnosis and aggressive surgical resection combined with irradiation.

In the present case, ACC demonstrates its striking propensity for perineural invasion in a very unique way along with the anecdotal site of spinal metastasis. Though the patient had a margin negative resection he did have a delayed perineural invasion in the form of an extremely rare, thoracic vertebral metastatic deposit at an extradural site. Intradural metastasis are common, however this case again demonstrates the rarity and need for high clinical suspicion to diagnose metastasis to an extradural spinal site in a margin negative resection for a Stage IVA disease (T2N2BM0, size of lesion 2.5 X2.3, 6 positive lymphnodes) of submandibular ACC.

## CONCLUSION

To conclude, early clinical suspicion in a young patient with proven primary in the salivary gland with sudden onset, progressive low back pain, mandates prompt radiological investigation in the form of an MRI of the affected vertebral segment and early surgical decompression/excision of the lesion.

Our case demonstrates the recurrence and intraspinal metastasis, after nine years of diagnosis, of an ACC of the submandibular gland. The extradural location of the metastatic thoracic vertebral deposit mandates its rarity and anecdotal value.

## REFERENCES


[1] Bohannon R, et al. Interrater reliability of a Modified Ashworth Scale of muscle spasticity. *Physical Therapy*. 1987;67:206-07.
[2] S Mishra, S lata, V kumar, G Mishra, P Ezhilarasu. Difficult Intubation; Temporomandibular Joint Ankylosis With Limited Mouth Opening And Hypertrophied Adenoid In A Six Year Old Child- Case Report And Review. The *Internet Journal of Anesthesiology*. 2008;22(1).
[3] Riela AR, Meyer D, McCool JA, Pilcula L. Metastatic-adenoid cystic carcinoma of the major salivary glands presenting as a spinal cord tumour. *Surg Neurol*. 1983;19(4):365–68.
[4] Trilling GM, Cho H, Ugas MA. Spinal metastasis in head and neck cancer. *Head & Neck Oncology.* 2012;4:36.
[5] Kazumoto K, Hayase N, Kurosumi M, Kishi K, Uki J, Takeda F. Multiple brain metastases from adenoid cystic carcinoma of the parotid gland. Case report and review of the literature. *Surg Neurol.* 1998;50(5): 475–79.
[6] Ko YH, Lee MA, Hong YS. Prognostic factors affecting the clinical outcome of adenoid cystic carcinoma of the head and neck. *Jpn J Clin Oncol.* 2007;37(11) 805–11.
[7] Perrin RG, Laperriere J, Loblaw A, Laxton AW. Spinal axis metastases. *Cancer metastatic to the central nervous system.* 2002;14:341-61.
[8] Simpson JR, Matsuba HM, Thawley SE, et al. Improved treatment of salivary adenocarcinomas: planned combined surgery and irradiation. *Laryngoscope*.1986;96:904-07.
[9] Harrison LB, Armstrong JG, Spiro RH, et al. Postoperative radiation therapy for major salivary gland malignancies. *J Surg Oncol*. 1990;45:52-55.
[10] Allen MS, Marsh WL. Lymph node involvement by direct extension in adenoid cystic carcinoma: absence of classic embolic lymph node metastasis. *Cancer*. 1976;38:2017-21.
[11] Carrasco Ortiz D, Aldape Barrios B. Adenoid cystic carcinoma of the dorsum of the tongue: presentation of a case (in Spanish). *Med Oral Patol Oral Cir Bucal.* 2006;11(5): E417-20.
[12] Tincani AJ, Del Negro A, Araújo PP, Akashi HK, Martins AS, Altemani AM, et al. Barreto G: Management of salivary gland adenoid cystic carcinoma: institutional experience of a case series. *Sao Paulo Med J.* 2006;124(1): 26-30.



**PARTICULARS OF CONTRIBUTORS:**
1. Assistant Professor, Department of Neurosurgery, Kasturba Hospital, Manipal, India.
2. Registrar, Kasturba Hospital, Manipal, India.
3. Assistant Professor, Department of Pathology, Kasturba Hospital, Manipal, India.
4. Assistant Professor, Department of Pathology, Kasturba Hospital, Manipal, India.
5. Professor and HOD, Department of Neurosurgery, Kasturba Hospital, Manipal, India.

**NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:**
Dr. Rajesh Nair,
Flat 505, Mandavi Paradise, Vidyarathna Nagr, Manipal-576104, India.
E-mail: rajeshnair39@yahoo.com



**FINANCIAL OR OTHER COMPETING INTERESTS:** None.

Date of Submission: **Jul 21, 2014**
Date of Peer Review: **Dec 04, 2014**
Date of Acceptance: **Dec 11, 2014**
Date of Publishing: **Jan 01, 2015**