Summer 3-24-2022

Radiological review of dumbbell-shaped spinal schwannoma with extradural and intradural-extramedullary components

Dr Sai Siddartha Kosinepalli
JSSMC

Dr. Sudha Kiran Das
JSSMC

Dr Gurumurthy B
JSSMC

TITILE: Radiological review of dumbbell-shaped spinal schwannoma with extradural and intradural-extramedullary components

HISTORY: C/O paresthesia in the bilateral lower limbs. No e/o pain. CBC, RFT, LFT, ESR, CRP are within normal limits.

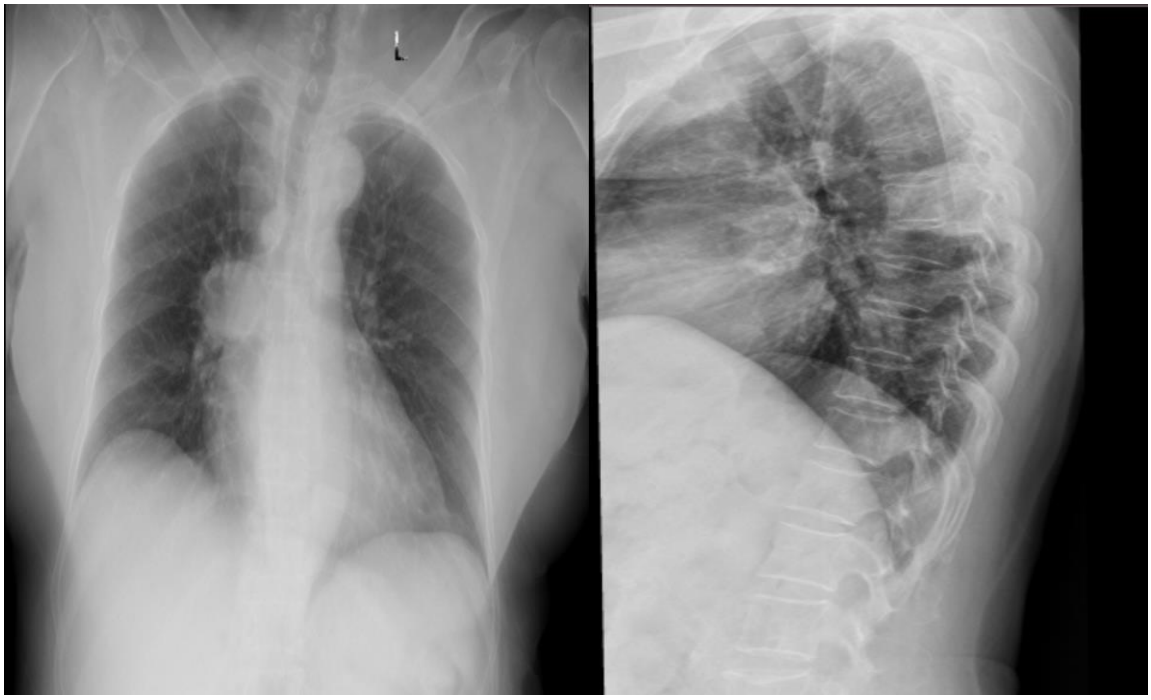

Figure 1A- Frontal chest radiograph shows a relatively well defined homogenous radiodense mass lesion in the right hilar region with indistinct medial border. The lesion is seen causing right widening of right paravertebral stripe with positive hilum overlay sign. Figure 1B- LATERAL DORSAL SPINE radiograph - Shows well defined homogenous mass lesion at D5-D6 vertebral level with indistinct posterior margin. The lesion is seen causing widening of neural foramina.

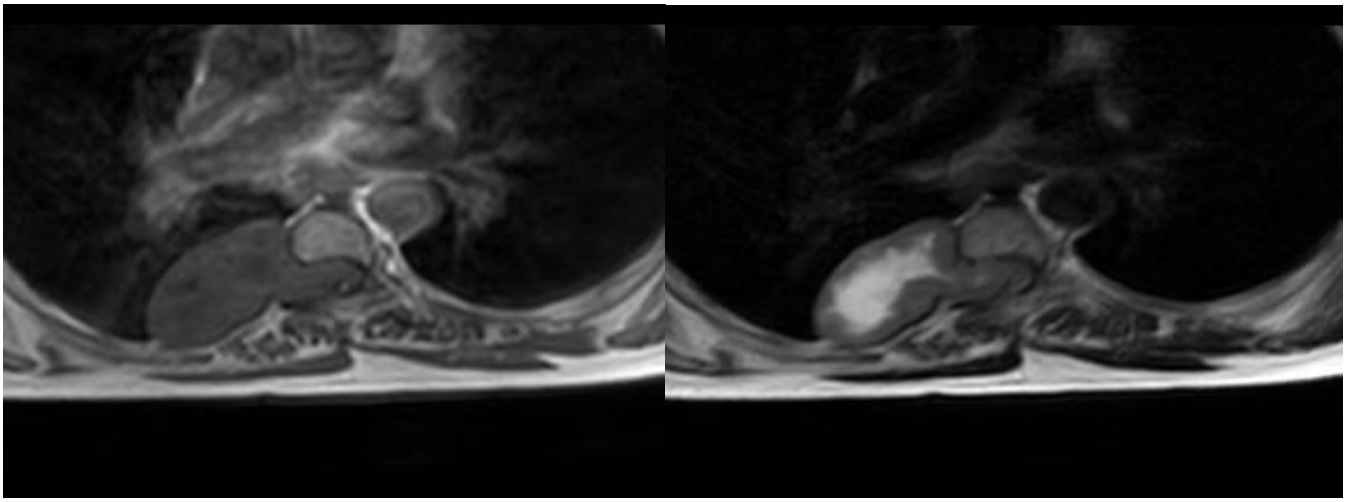

Figure 2 a & b - MRI T1W & T2W AXIAL SECTIONS - well-defined dumb-bell shaped intradural extramedullary mass lesion at the D5-D6 intervertebral level on the right side with extraforminal extension causing right neural foraminal widening appearing peripheral T2 hypointensity & central T2 hyperintensity and isointense on T1.

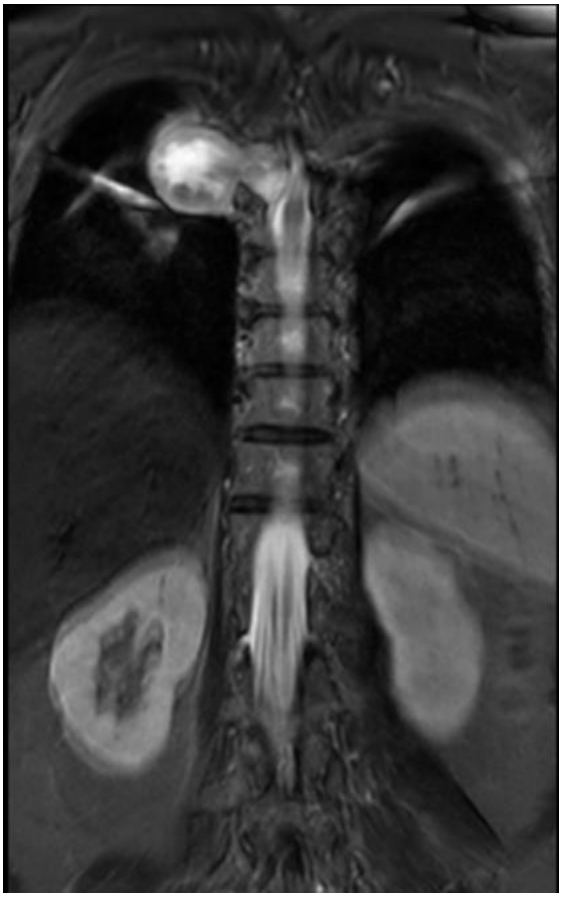

Figure 3- MRI STIR CORONAL SECTION – Shows peripheral hypointensity & central STIR hyperintensity.

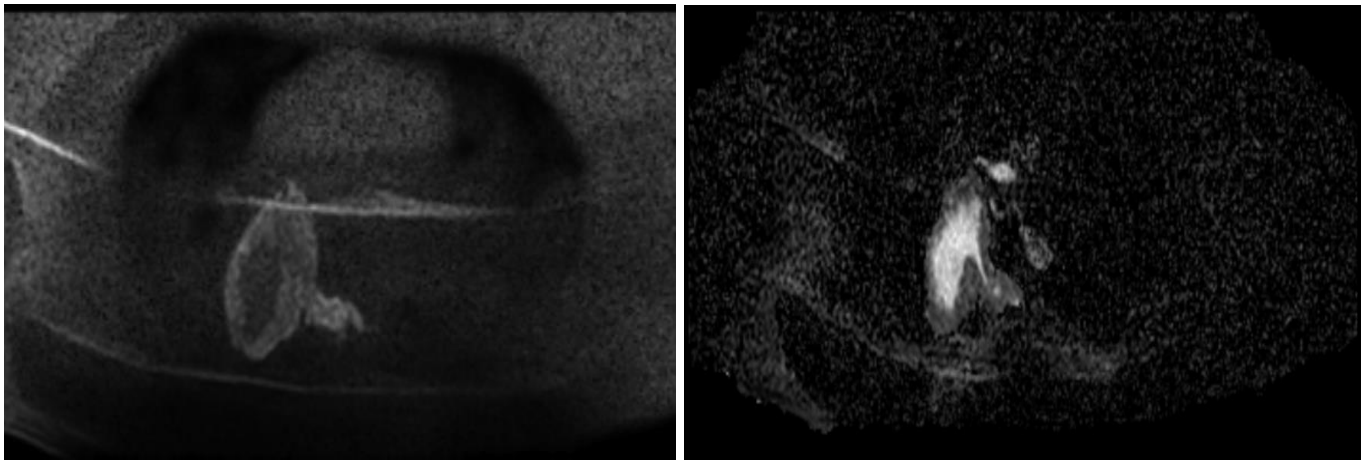

Figure 4- DWI AND ADC – Shows no restriction diffusion

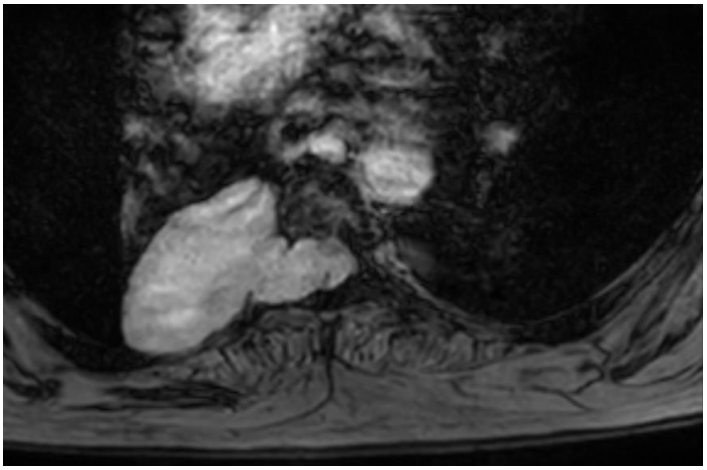

Figure 5- FFE SEQUENCE- shows no blooming.

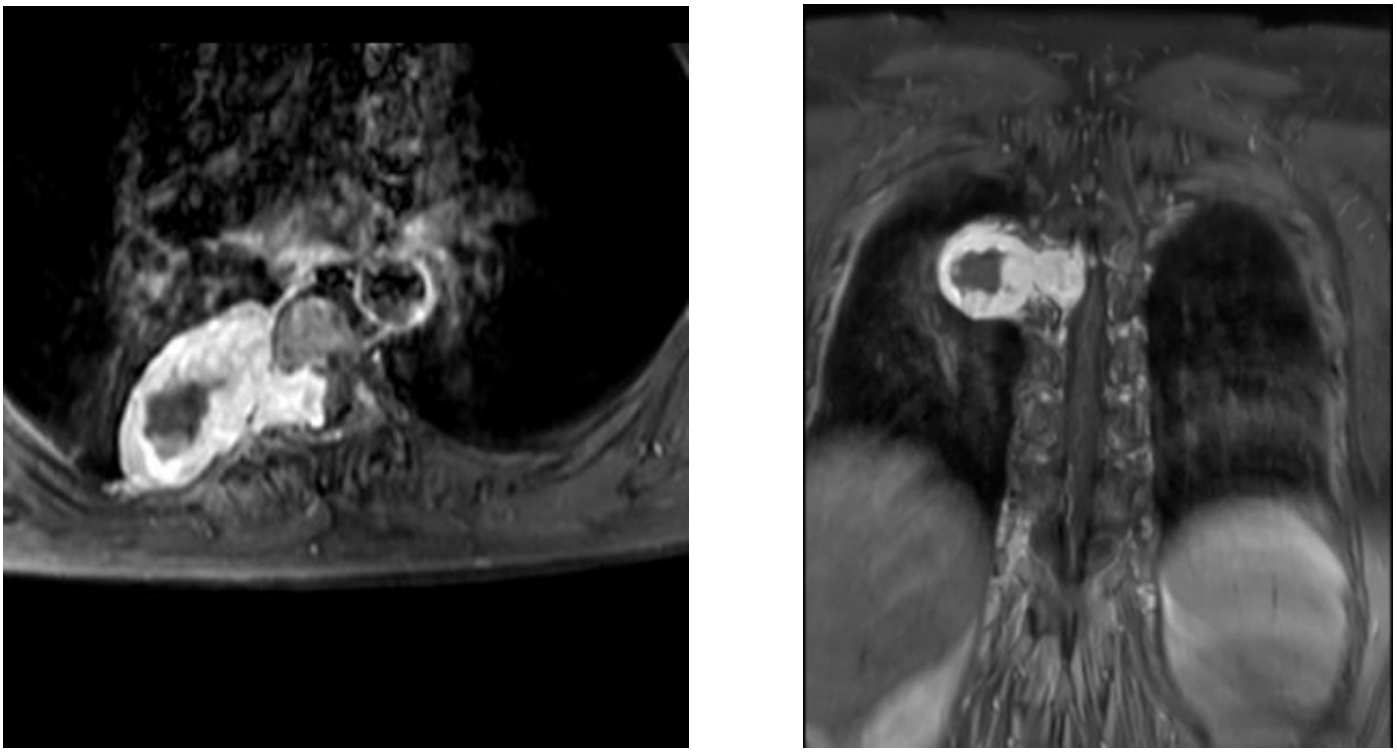

Figure 6 A & B - MRI CONTRAST T1W AXIAL & POSTCONTRAST T1W CORONAL SECTIONS – On postcontrast images the lesion is seen heterogeneously enhancing with central non-enhancing necrotic areas and peripheral rim of heterogeneous enhancement.

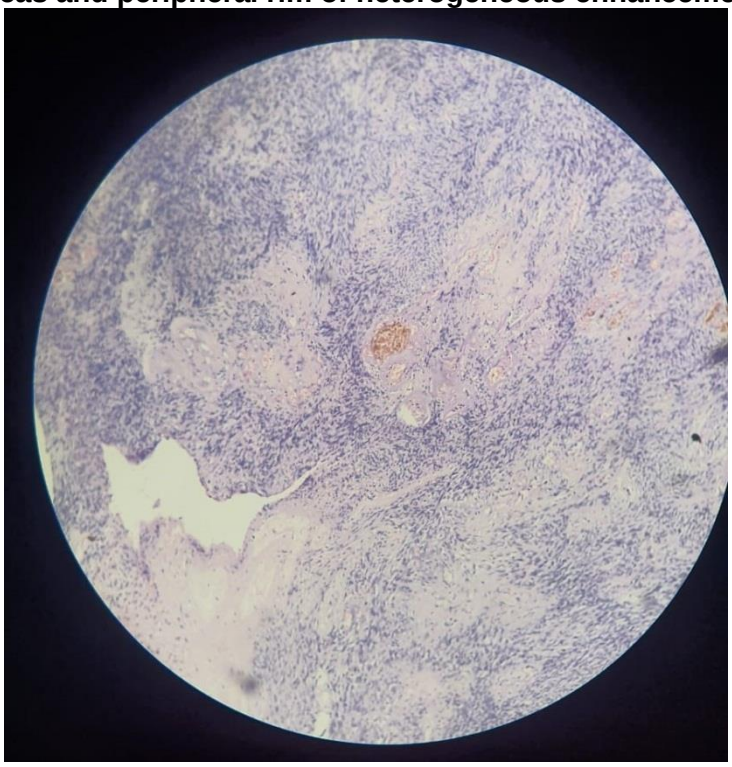

Figure 7 – HISTOPATHOLOGY- Shows hypercellular Antoni A and hypocellular Antoni B cells with Verocay bodies.

DIFFERENTIAL DIAGNOSIS: Peripheral nerve sheath tumor and neurogenic tumor.

FINAL DIAGNOSIS: Well defined dumbbell shaped peripherally enhancing intradural extramedullary mass lesion at the D5-D6 intervertebral level with right neural foraminal narrowing– Schwannoma.

HISTOPATHOLOGY: Showed hypercellular Antoni A and hypocellular Antoni B cells with Verocay bodies. The features are suggestive of schwannoma.

DISCUSSION:

Schwannomas are among the most common of the peripheral nerve sheath tumors. These are the most common intradural extramedullary spinal tumors, accounting for 15-50% of spinal lesions. Schwannomas are seen after 5th decade and peaks in 7th decade. Schwannomas are associated with neurofibromatosis 2 (NF-2).

ETHIOPATHOGENESIS:

Schwannomas arise from Schwann cells. These cells produce an insulating sheath surrounding the nerve fibers. The Schwann cells manufacture the nerve sheath out of myelin. Schwann cells doesn't exist in the spinal cord. They produce their myelin sheath only for nerves outside the brain and spinal cord, called peripheral nerves.

Spinal schwannomas do not arise in the spinal cord as there are no Schwann cells. Instead, schwannomas arise on the spinal nerve roots.

IMAGING CONSIDERATION:

CHEST RADIOGRAPHY -

1. The posterior mediastinal mass will cause obliteration or widening of paravertebral stripe without silhouetting cardiac border with hilum overlay sign.
2. The "sulcus" sign is seen on lateral radiograph. This sign is seen in peripheral nerve sheath and neurogenic tumor. Whereas ganglionic cell tumor does not have sulcus.

MR IMAGING FEATURES –

- ✓ T1 weighted images shows intermediate to low signal intensity.
- ✓ T2 weighted images show heterogeneously high signal intensity.
- ✓ T2* weighted images show areas of hemosiderin.
- ✓ T1 contrast (gadolinium) shows heterogeneously enhancement. Target sign is also seen in postcontrast images with peripheral enhancement and central non enhancement.
- ✓ Split-fat sign: Thin peripheral rim of fat best seen on planes along long axis of the lesion in non-fat-suppressed sequences.
- ✓ Fascicular sign: multiple small ring-like structures.

- ✓ "Target sign" is seen on T2W appearing central hypointensity and peripheral hyperintensity. Peripheral hyperintensity is due to histological variation with a dense central area of collagenous stroma surrounded by myxomatous tissue.

Treatment: Schwannomas are slow-growing lesions. Surgery is the treatment of choice. As schwannomas do not infiltrate the parent nerve, they can usually be separated from it. Recurrence is unusual, after complete resection. They almost never undergo malignant change.

TEACHING POINTS:

- ✓ Spine schwannoma is the dumbbell shaped extramedullary intradural benign tumor causing neural foraminal narrowing and without causing bone remodeling. The target sign, fascicular sign and split- fat sign are characteristic signs of schwannoma on MRI.
- ✓ Schwannoma appears as a posterior mediastinal mass on frontal radiograph causing widening or obliteration of paravertebral stripe with sulcus sign on lateral chest radiograph.

References:

i. Kurtkaya-Yapicier O, Scheithauer B, Woodruff JM. The pathobiologic spectrum of Schwannomas. Histol. Histopathol. 2004;18 (3): 925-34.
ii. Murphey MD, Smith WS, Smith SE et-al. From the archives of the AFIP. Imaging of musculoskeletal neurogenic tumors: radiologic-pathologic correlation. Radiographics. 19 (5): 1253-80.
iii. Osborn AG. Diagnostic neuroradiology. Mosby Inc. (1994) ISBN:0801674867
iv. Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic-pathologic correlation. Radiographics. 24 (5): 1477-81.
v. Louis DN, Ohgaki H, Wiestler OD et-al. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol. 2007;114 (2): 97-109.