

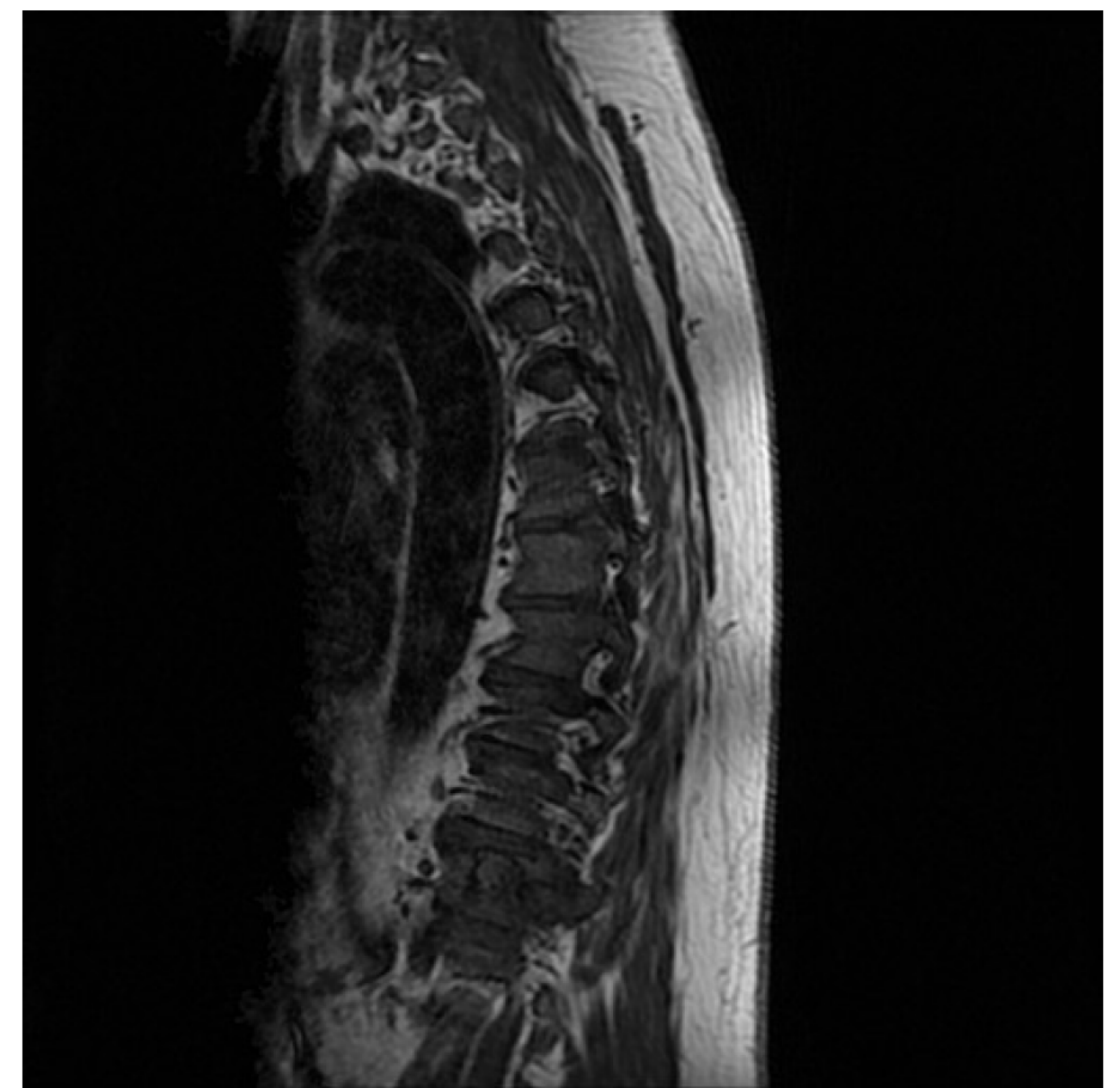
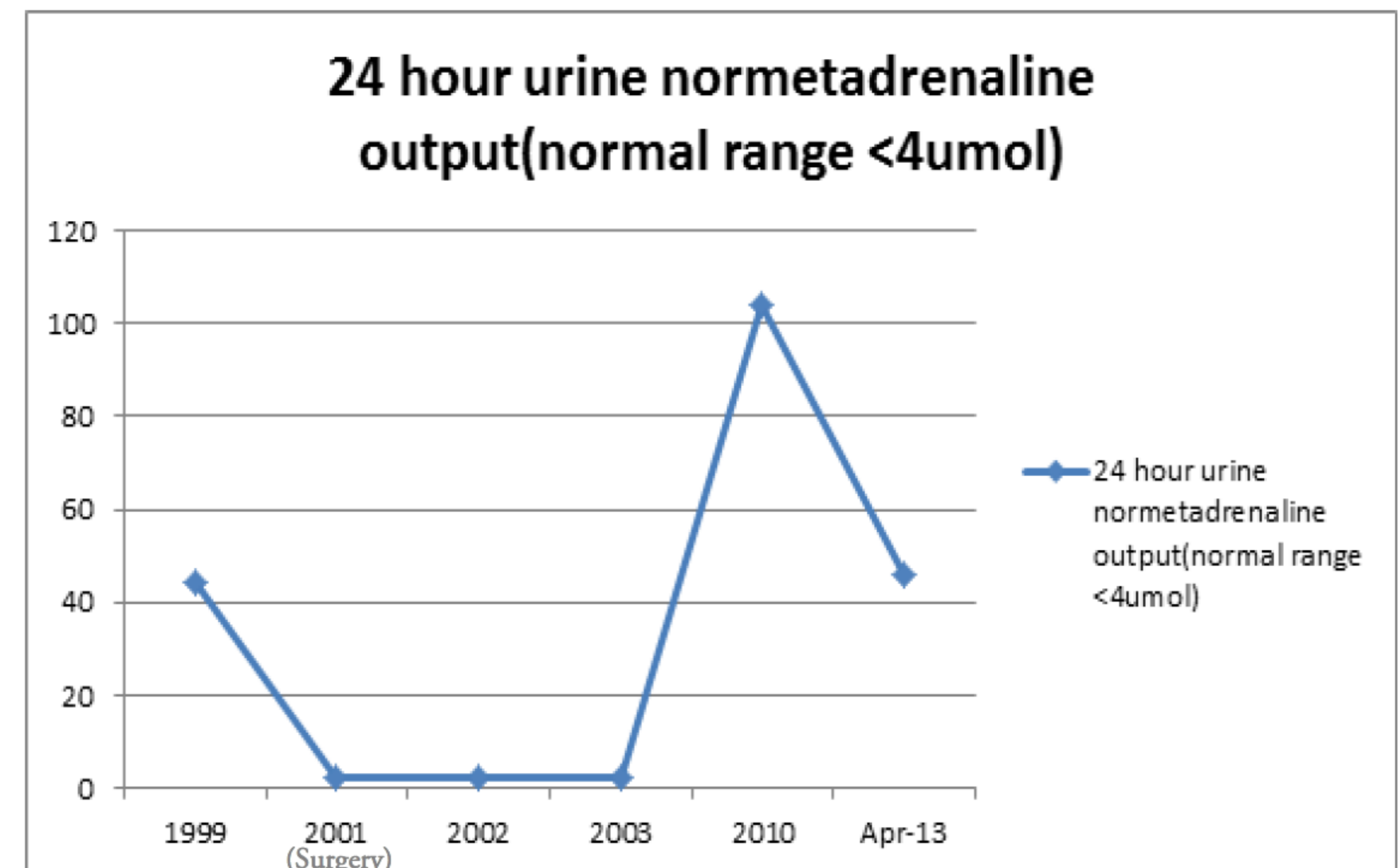
## Case History

A 50 year old man presented with pain abdomen. Ultrasound scan of abdomen showed a right adrenal mass which measured 7.5 cm on CT scan. Urine 24 hour catecholamines were raised to ten times above normal. MIBG scan was consistent with a Pheochromocytoma which was then resected and postop urine catecholamines returned to normal. Genetic testing for VHL, MEN2 and NF1 was negative. Patient was discharged from endocrine clinic after four years of annual follow up during which urine catecholamines remained normal. He presented to casualty 7 years post discharge from clinic with 3 months history of progressive back pain and recent leg weakness. MRI scan showed D12 collapse with cord compression. Urgent spinal fixation was done. MIBG scan showed a lesion consistent with Pheochromocytoma which was confirmed on biopsy. A prolonged course of radiotherapy led to full recovery of function.

He has developed paraparesis after a further five years and C3-4 metastatic compression has been found. Urine 24 hour catecholamines are significantly raised. CT scan of abdomen is normal. Chromogranin B is slightly raised. Tumour debulking and radiotherapy has been done with gradual recovery of mobility. Further therapeutic MIBG was deemed unsafe. Patient is due to start treatment with Sunitinib.

## Discussion

At least ten percent of Pheochromocytomas and up to twenty-five percent of extra-adrenal abdominal and mediastinal secretory paragangliomas are malignant. Clinical, Histopathological or biochemical features do not predict malignant behavior in case of Pheochromocytoma. In case of paragangliomas, those with succinate dehydrogenase subunit B gene often metastasize. Surgery helps with tumor debulking to relieve pressure effects and to reduce catecholamine output and alleviate symptoms. Chemotherapy with cyclophosphamide, vincristine and dacarbazine with or without doxorubicin has been used with variable success. Molecular targeted therapies including Sunitinib, a tyrosine kinase inhibitor have shown promise in early trials and more results are awaited.



MRI showing T12 vertebral body collapse with a soft tissue mass anterior and posterior to this vertebral body and extending into the spinal canal.

## Conclusion

Current guidelines for duration of post operative follow up in case of solitary non familial Pheochromocytoma vary but we recommend life long follow up with annual urine catecholamine measurement.

