A giant symptomatic neurinoma located in the pelvi: a case report.

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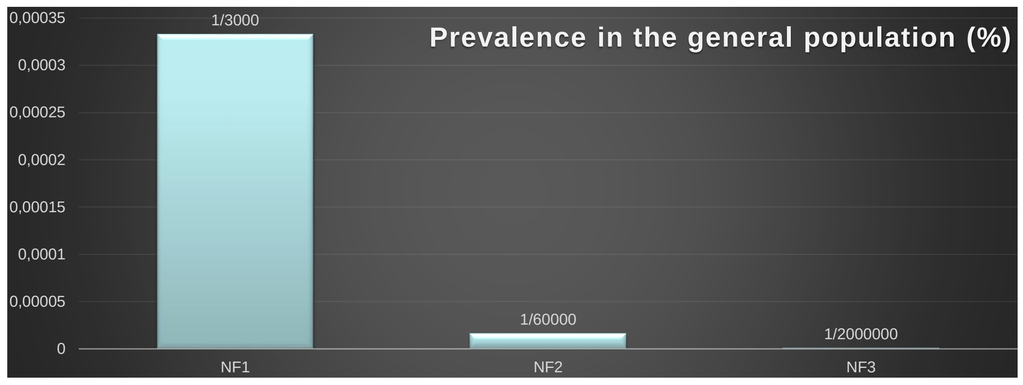
INTRODUCTION

Neurofibromatosis is an inherited neurocutaneous condition that increases the risk of tumor formation. It can be divided into three major subtypes: neurofibromatosis 1 (NF1), neurofibromatosis 2 (NF2) and schwannomatosis (NF3). Clinical features of NF1 include neurofibromas, café-au-lait lesions, freckling and optic glioma. NF2 is characterized by bilateral vestibular schwannomas and other CNS tumors. NF3 is characterized by multiple schwannomas without the involvement of the vestibular nerves and usually associated with pain

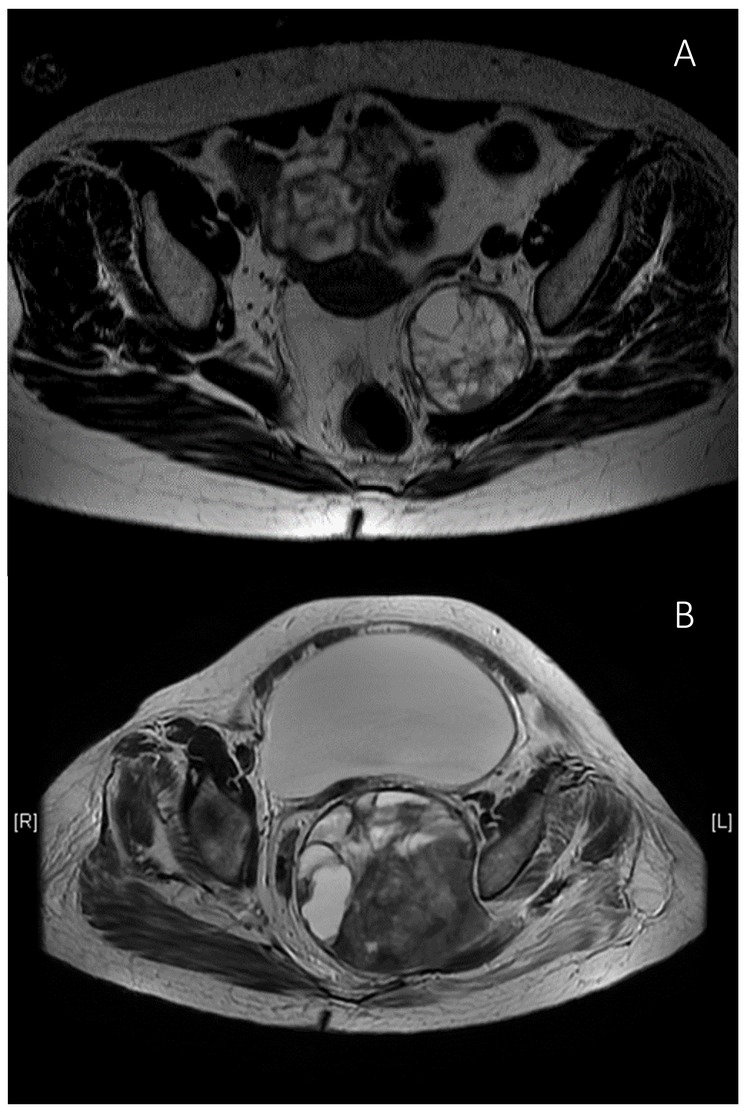
	NF1	**NF2**	**NF3**
Gene (inheritance)	NF1 (*AD*)	NF2 (*AD*)	SMARCB1/INI1 (*AD*)



CASE REPORT

We want to describe an unusual presentation of neurinoma that caused an important mass effect. Our patient (C.P. 69 years old - female) is suffering from neurofibromatosis since her young age with a negative genetic test result. She has undergone many surgical excision of the neurofibromas during her life, mainly in the spine. She has not cutaneous stigmate of NF1 neither Lish nodules.

She was admitted to our hospital for chronic pain in the left leg that she described as the worst possible, a mild weakness of the ipsilateral distal limb, antalgic gait and constipation. Subsequent MRI showed multiple neurinomas localized near the seventh rib, along the spine, between the psoas and the iliacus muscle and in the pelvis. The last one tumor was 10 x 10 x 8.7 cm in size and compress and displaced the rectum to the right side. Her pain was treated with pregabalin BID and she underwent a combined abdominal surgery and neurosurgery. The outcome was good but the mass was not completely removed. The post-surgery recovery was complicated by delirium which is lasted a few days. At the discharge the pain was relieved.



Axial T2-weighted images show the pelvic neurinoma.
(A) It was obtained in 2009 (B) It was obtained in 2016. The size was remarkably increased.

CONCLUSIONS

Pelvic location of neurinomas is uncommon and there are few case reports in literature. It is important to remember that neurofibromatosis can manifest with many symptoms and physicians should pay attention to unexpected rare presentation. Furthermore it is important to emphasize that a multidisciplinary team approach is the better way to treat this disease.

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