Report of a spinal ganglioneuroma case (Intra- & extradural tumor) in a 23 years old patient at Fatemi Hospital of Ardabil, Iran

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Misagh Pourdonya[1], Mehdi Chiniforoush[2]

[1]Medical Student, Ardabil University of Medical Sciences

[2]Pathology Assistant Professor, Anatomical Sciences Group, Ardabil University of Medical Sciences

INTRODUCTION

Ganglioneuromas are rare, slow growing, benign tumors that generally arise from the ganglion cells of the sympathetic chain, but they may also arise from sympathetic nerves as well as from peripheral nerves. They represent the most benign form of neurogenic tumor with 60% of them occurring in children and young adults (2, 3, 4, 5, 6). The ratio of male to females is approximately 3:2. They occasionally grow to large size but total excision using microsurgical techniques is often possible, and may be curative.

Ganglioneuromas are considered, as a rule, benign tumors. However, in exceptional cases, intra-tumoral areas of malignant transformation, metastasis, and the development of malignant peripheral tumors arising from ganglioneuromas have been described (7,8,9,10,11). Although they usually occur in relation to the adrenal medulla and sympathetic chain in the retroperitoneal and retropleural spaces, they may be also be found along in the intestinal tract, and occasionally in a peripheral nerve. Multiple locations are possible. They have also been described in association with Neurofibromatosis 1 diseases (12,13).

CASE REPORT

The patient S.B is a 23 year old man who hospitalized on 30 August 2014 at males' neurosurgery ward with right extremities hemiparesis which had been started for one year. After clinical and neurological examinations, the first diagnosis, Cervicoccipital tumor was proposed. Then for the final diagnosis MRI & CT scan were recommended (Fig 3-6). Having diagnosed a vast extra axial tumor at the C1 & C2 levels at MRI, Neurinoma was considered. His laboratory tests revealed the patient had leukocytosis (20.2 per microliter) with an erythrocyte count of 4.08 permicroliter and hematocrit of 36.3%. Other factors were about normal. Later that day the patient underwent a laminectomy operation on his C1 & C2 levels and the tumor of that region which concurrently was intra- and extradural completely was evacuated. After operation on 1 September 2014 he had a suitable general condition, sensory and motor function tests of extremities and his right side hemiparesis were considerable improved. Observing smooth tissue containing paramorphic irregular gray components in macroscopic view, Ganglioneuroma at histopathologic study and microscopic view was diagnosed (Figure 1&2).

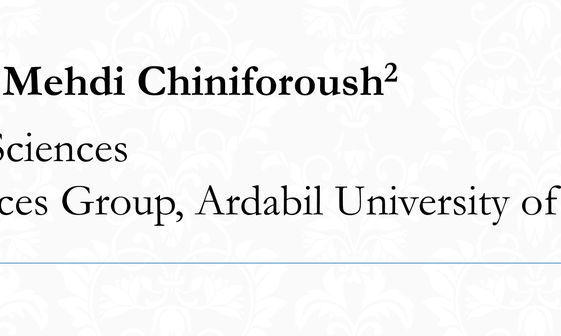

Fig. 3

Fig. 4

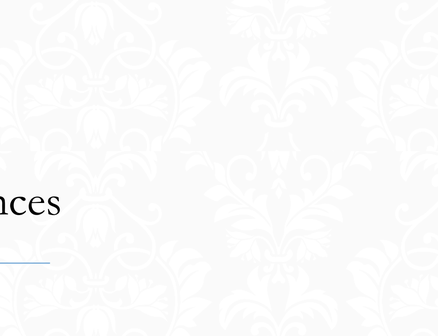
Fig. 1

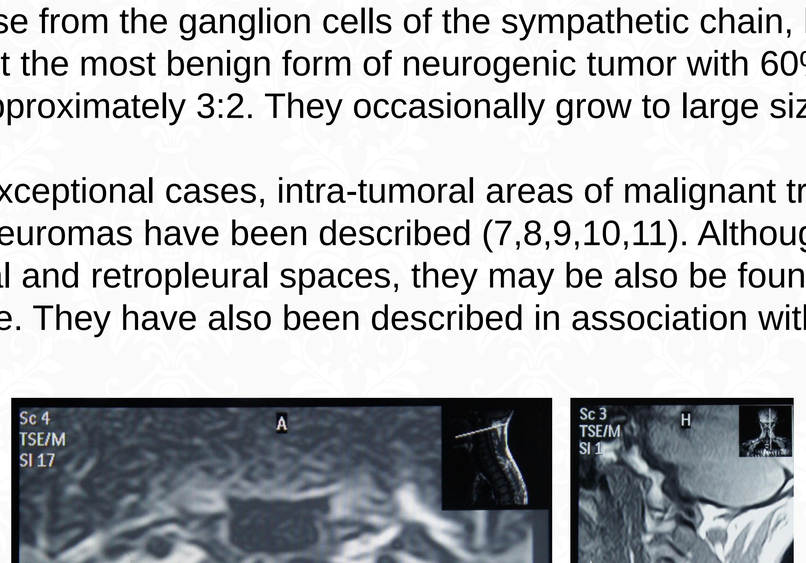

Fig. 5

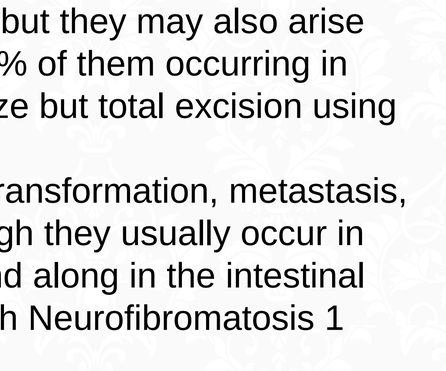
Fig. 2

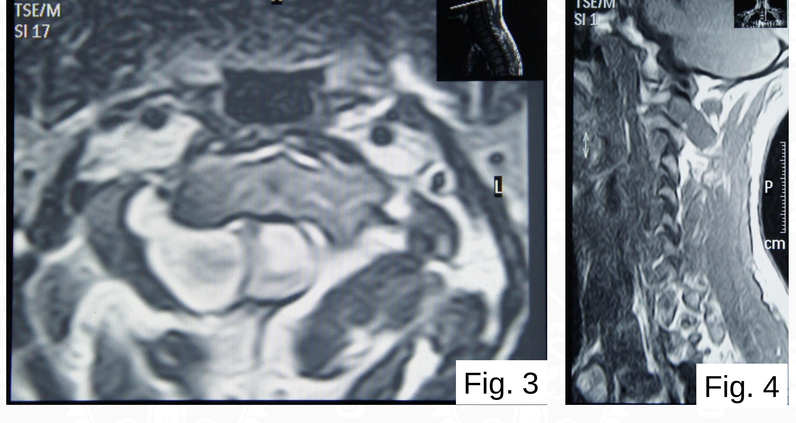

Fig. 6

DISCUSSION

Ganglioneuromas reside within a class of neuronal tumor in which the neoplastic cells express a mature neuronal phenotype (14). This type of tumor consists of well-differentiated large pyramidal-shaped ganglion cells embedded within a scanty sroma of spindle cells (15,16)(Figure 1). Ganglioneuromas are usually white, firm and encapsulated, slow-growing tumors. Microscopic examination reveals large ganglion cells and areas of smaller lymphocyte-like cells within a matrix of fibrous and Schwann cells (Figure 1). Multinucleate cells with a well-defined nucleolus in each nucleus are commonly found (5). As diagnosis of ganglioneuroma is based on the absence of necrosis or immature ganglion cells (2), the entire tumor must be examined to rule out areas of malignant transformation.

Although ganglioneuromas can produce symptoms related to the large volume they may attain, most of them are asymptomatic and can be diagnosed incidentally by palpation in cases of superficial location(2,17), or on radiographic studies(12).

In some series, 0.8 to 3.5% of ganglioneuromas were dumbbell tumors(1,5). It is also well known that ganglioneuromas at or near the cervical spine are extremely uncommon(18,19). Regarding treatment, complete excision is the best option (2). When there is spinal cord compression surgical decompression must be undertaken as soon as possible (5, 18). If a complete resection can be achieved, there is no evidence supporting the use of any adjuvant postoperative therapy in the management of ganglioneuromas (2,3,6,20).

CONCLUSION

Ganglioneuromas occurring within the spinal column are exceedingly rare and may grow to a large size. Despite this size and the common involvement of both intra- and extra-spinal compartments, favourable outcome, with good functional recovery is often possible after complete excision using microsurgical techniques.

References

1.Shepard RH, Sutton D. Dumbbell ganglioneoromata of the spine with a report of four cases. Br J Surg 1958;45:305-17.
2.Barthelemy I, Belveze P, Emering C, Reynaud P, Beaujard H, Peri G, Mondie JM. Cervical ganglioneuroma. Review, apropos of a case. Rev StomalChirMaxillofac 1998;99:210-13.
3.Belzberg AJ, Campbell JN. Neoplasms of peripheral nerves. In: Wilkins RH, Rengachary SS, ed. Neurosurgery 2nded. New York: McGraw-Hill, 1996:3217-23.
4.Fagan CJ, Swischuk LE. Dumbbell neuroblastoma or ganglioneuroma of the spinal canal. AJR 1974;120:453-60.
5.Miura Y, Okumichi T, Yoshioka K, Okumichi K, Kajihara H. Successful excision of a "dumb-bell" shaped ganglioneuroma of the posterior mediastinum with a large intraspinal component. Eur J Surg 1993;159:635-8.
6.Mutluer S, Ersahin Y, Binatli O, Demirtas E. Dumbbell ganglioneuroma in childhood. ChilNervSyst 1993;9:182-4.
7.Chandrasoma P, Shibata D, Radin R, Brown LP, Koss M. Malignant peripheral nerve sheath tumor arising in an adrenal ganglioneuroma in an adult male homosexual. Cancer 1986;57:2022-5.
8.Drago G, Pasquier D, Pinel N, Rouault-Plantaz V, Dyon JF, Durand C, Armari-Alla C, Plantaz D. malignant peripheral nerve sheath tumor arising in a "de novo" ganglioneuroma: a case report and review of the literature. Med PediatrOncol 1997;28:216-22.
9.Garvin H, Lack EE, Berenberg W, Frantz CN. Ganglioneuroma presenting with differentiated skeletal metastases. Cancer 1984;54:357-60.
10.Kulkarni AV, Bilbao JM, Cusimano MD, Muller PJ. Malignant transformation of ganglioneuroma into spinal neuroblastoma in an adult. Case report. J neurosurg 1998;88:324-7.
11.Ricci A, Parham DM, Woodruff M, Callihan T, Green A, Erlandson RA. Malignant nerve sheath tumours arising from ganglioneuroma. Am J SurgPathol 1984;8:19-29.
12.Geraci AP, de Csepel J, Shlasko E, Wallace SA. Ganglioneoroblastoma and ganglioneuroma in association with neurofibromatosis type 1: report of three cases. J Child Neurol 1998;13:356-8.
13.Suetake K, Niwa J, Okuyama T, Shimoyama N, Ishidate T. Ganglioneuroma in the cervical ganglion with neurofibromatosis-2: a case report. No ShinkeiGeka 1993;21:629-32.
14.Kleihues P, Burger PC, Scheithauer BW. Histological typing of Tumours of the Central Nervous System. WHO International Histological Classification of tumours. 2nded. New York: Springer Verlag; 1993.
15.Shimada H, Brodeur G. Tumors of peripheral neuroblasts and ganglion cells. In: Bigner D, McLendon R, Bruner J, editors. Russell and Rubinstein's Pathology of tumors of the Nervous system. New York: Oxford University Press; 1998. p. 493-533.
16.Stout A. Ganglioneuroma of the sympathetic nervous system. SurgGynecolObstet 1947;84:101-109.
17.RegasJs, Sanchez de toledoJ,MarquesGubern A, Tresserra L, Balcells R. Primary cervical tumors of the sympathetic nervous system. Report of 8 cases. Cir Pediatr 1989;2:86-9.
18.Maggi G, Dorato P, Trischitta V, Varone A, Civetta F. Cervical dumbbell ganglioneuroma in an eighteen-month old child. A case report. J NeurosurgSci 1995;39:257-60.
19.Shotton JC, Milton CM, Allen JP. Multiple ganglioneuroma of the neck. J LaryngolOtol 1992;106:277-8.
20.Campbell R. Tumors of peripheral and sympathetic nerves. In: Youmans JR, ed. Neurological Surgery 3rd ed. Philadelphia: WB Saunders Co 1990:3667-75.