BACKGROUND

Ganglioneuroblastomas are uncommon primitive neuroectodermal tumors (PNETs) reported almost exclusively in childhood. They are composed of a mixture of mature gangliocytes and immature neuroblasts and are considered a neoplasm with intermediate malignant potential between ganglioneuroma and neuroblastoma. Since their origin from primordial neural crest cells they usually arise where sympathetic tissue is located, mainly in retroperitoneum and mediastinum, while their presentation in central nervous system is uncommon and intraspinal location is even rarer.

CASE REPORT

A 1 year-old boy presented with persistent stiff neck and head flexion to the left side. A RM image showed an intra-axial mass extended from C5 to D5, partially cystic and partially solid, with an intense contrast enhancement, diagnosed as first hypothesis as ependymoma. The patient was submitted to laminectomy at C7 and T2 level for neoplasm excision. No residual tumor was seen at post-surgical RMI. Histological examination revealed a population of immature neuroblasts, synaptophysin-positive, with focally brisk mitotic activity, admixed with nests of mature ganglion cells, focally positive for NeuN. The final diagnosis was ganglioneuroblastoma, grade IV according to WHO classification 2007. The patient recovered completely from neurological symptoms and was submitted to high-doses chemotherapy and protocol intensification with autologous stem cells transplant. No disease recurrence was documented 2 years later.

CONCLUSIONS

Ganglioneuroblastomas arising in thoracic spine are extremely rare neoplasms and only few case are reported. The standard treatment for ganglioneuroblastoma is the complete surgical removal and adjuvant radiotherapy is recommended in adults, while chemotherapy showed various results. As spinal cord is an unusual localization of these tumors it is hard to predict their behavior and their response to post-surgical treatments. Moreover there are no specific radiological features and pathologists should be aware of this entity to be considered in differential diagnosis of spinal cord pediatric tumors.