Coexisting Neurofibromatosis-II And Sarcoidosis: A Case Report

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Abstract

We report about a young woman who had various biopsies in the cervical region because of cervical swelling. Histology showed neurofibromas as well as sarcoidosis. MRI revealed bilateral schwannoma.

CASE REPORT

We report about a 30-year-old woman who had burning pain in the cervical region. There were enlarged and tender lymph nodes especially in the submandibular angle bilaterally. The patient had a history of prior lung sarcoidosis two years ago which was successfully treated with steroids. This time chest-CT showed no pathological changes.

The total white blood count was in normal range (6150/ L) but showed increased neutrophiles (4630/ L), increased monocytes (590/ L), decreased lymphocytes (1060/ L) and decreased eosinophiles. Red blood count and platelets were in normal range. Serum ACE was slightly elevated (56 IU/L). Blood chemistry showed low total iron of 33/ g. Three specimens, which were clinically suspected to be lymph nodes (two from the neck and one from the upper arm) were taken.

The Histology showed Schwannomma with no signs of malignancy and no signs of sarcoidosis (fig.2). However, histology of submandibular lymph nodes a few months before showed typical histomorphology of sarcoid granulomas (figure 1).

Figure 1

Figure 1: Non necrotizing granuloma formation in Sarcoidosis (hemotoxylin-eosin stain)

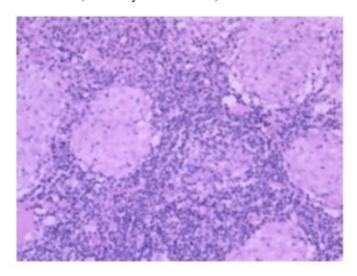
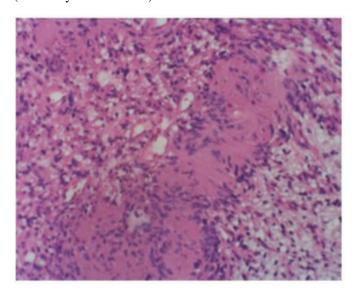


Figure 2

Figure 2: Palisading of fibroblast like cells in Schwannoma (hemotoxylin-eosin stain)



On MRI the patient had bilateral lymph node enlargement (figure 4) and bilateral Schwannoma (dumbell-tumor) of the nerve roots C7 (figure 3 arrow).

Figure 3

Figure 3: CM-enhanced coronal T1w image shows bilateral Schwannoma with slight contrast enhancement at the level of C7

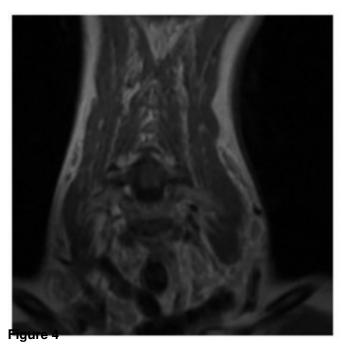
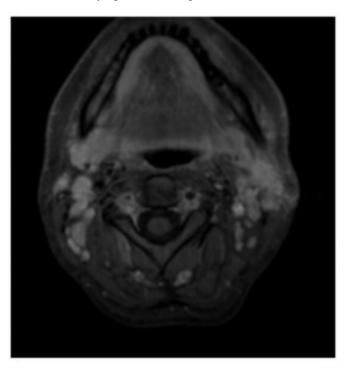


Figure 4: CM-enhanced and fatsaturated axial T1w image

shows bilateral lymph node enlargement of sarcoidosis



DISCUSSION

Bilateral Schwannoma is one of the typical features of NF-2 [1]. In addition the patient had sparse manifestations of cutaneous neurofibromas. Other typical signs of Neurofibromatosis-2 such as acoustic schwannomas, meningeomas or other intracranial tumours (especially schwannomas of the cranial nerves) were rouled out by MRI.

Sarcoidosis is a systemic noncaseating granulomatous disorder of unknown origin. The prevalence of sarcoidosis in Europe is 19/100,000 and involvement of lymph nodes is common (75-90%) [$_2$].

To our knowledge coincidence of NF-2 and sarcoidosis has not been described yet (Medline search).

References

- 1. Akeson P, Holtas S: Radiological investigation of neurofibromatosis type 2, Neuroradiology 36: 107-110, 1994.
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