AJNR

Isolated Castleman disease of the neck: MR findings.

M Glazer, V M Rao, D Reiter and P McCue

AJNR Am J Neuroradiol 1995, 16 (4) 669-671
http://www.ajnr.org/content/16/4/669

Isolated Castleman Disease of the Neck: MR Findings

Mark Glazer, Vijay M. Rao, David Reiter, and Peter McCue

Summary: Castleman disease in an 11-year-old girl appeared as a neck mass that grew despite antibiotic treatment. MR showed a well-defined solid mass, isointense with muscle on short-repetition-time/short-echo-time images, with a stellate area of central hypointensity on long-repetition-time/long-echo-time images, that did not enhance with gadolinium.

Index terms: Neck, neoplasms; Children, neoplasms

Castleman disease is a benign lymphoid neoplasm that usually is found in the mediastinum. Isolated Castleman disease in the neck is rare; cervical lymph node enlargement may be the only clinical presentation of this disease. We present a case with magnetic resonance (MR) findings of cervical Castleman disease.

Case Report

An 11-year-old Italian girl presented with a right neck mass that grew slowly over 3 months despite treatment with multiple antibiotics. She had no significant medical history and was afebrile. The mass was palpable on the right just deep to the middle third of the sternocleidomastoid muscle. Initial ultrasound examination revealed a 3.2 $\times$ 2.5 $\times$ 1.6-cm ovoid mass that had the appearance of an enlarged lymph node. Results of her mononucleosis test (heterophile test) and Epstein Barr virus antibody tests were negative. She had a normal complete blood cell profile.

MR examination of the neck was performed on a 1.5-T magnet (GE Signa, Milwaukee, Wis) with short-repetition-time/short-echo-time images in sagittal and axial planes, an axial fast spin-echo, long-repetition-time/long-echo-time sequence, and coronal and axial short-repetition-time/short-echo-time sequences after gadopentetate dimeglumine administration. A well-defined solid mass in the posterior triangle of the right neck was noted (Fig 1A), beginning at the level of the hyoid bone and extending to the level of the thyroid gland. It was isointense with muscle on the short-repetition-time/short-echo-time images. There was a stellate area of central hypointensity noted on the long-repetition-time/long-echo-time images (Fig 1B). Postgadolinium images demonstrated no evidence of enhancement (Fig 1C). The patient underwent excision of the right neck mass. Histopathologic evaluation revealed giant lymph node hyperplasia (Castleman disease), hyaline-vascular type (Fig 1D). The central hilar region of the node showed marked sinus histiocytes and radial fibrosis (Fig 1E). Subsequent chest and abdomen MR findings were normal.

Discussion

In 1956, Dr Benjamin Castleman described 13 patients with a localized mediastinal lymph node hyperplasia resembling thymoma. This entity currently is termed *localized nodal hyperplasia*, *angiomatous lymphoid hamartoma*, *giant lymph node hyperplasia*, or *Castleman disease* (1).

Keller's group (2) classified 81 cases of giant lymph node hyperplasia into two histologic types: the hyaline vascular type, which comprised 91% of their lesions; and the less common plasma cell variant. The hyaline vascular type consists of large fibrous masses in the perivascular area with interspersed areas of plasma cells. These hyalinized capillaries are surrounded by lymphocytes. Most lesions in the neck are of this type, as was the lesion in our case. These patients usually are asymptomatic except for complaints related to the local mass effect (2–11).

The plasma cell variant has sheets of mature plasma cells in the interfollicular tissue with small areas of hyaline (2, 8). In approximately 50% of these cases, patients demonstrate systemic signs and symptoms, including fever, elevated sedimentation rate, and hypergammaglobulinemia (12).

Received December 8, 1992; accepted after revision June 17, 1993.

From the Department of Radiology (M.G., V.M.R.), the Center for Plastic Surgery (D.R.), and the Department of Pathology and Cell Biology (P.M.), Thomas Jefferson University Hospital and Jefferson Medical College, Philadelphia, Pa.

Address reprint requests to Vijay M. Rao, MD, Department of Radiology, Thomas Jefferson University Hospital and Jefferson Medical College, 1092 Main Bldg, 132 S 10th St, Philadelphia, PA 19107.

AJNR 16:669–671, Apr 1995 0195-6108/95/1604–0669

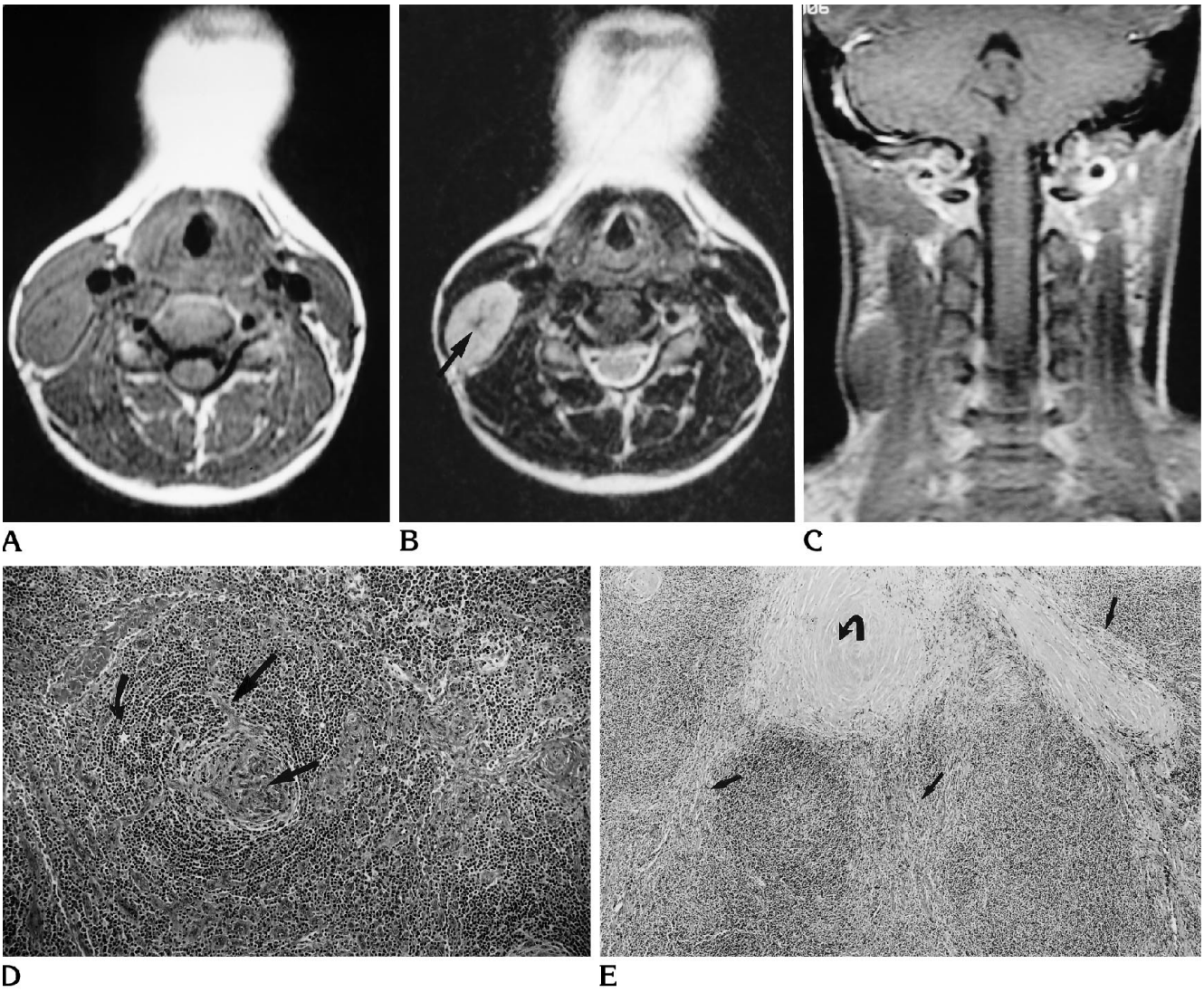

Fig 1. An 11-year-old girl with an enlarging mass on the right side of her neck.

A, Precontrast T1-weighted (450/12/2 [repetition time/echo time/excitations]) axial image shows a well-circumscribed mass isointense with muscle in the right posterior triangle of the neck.

B, Precontrast T2-weighted, fast spin-echo (9500/84/1) axial image shows the mass to be hyperintense relative to muscle. Note the central stellate hypointensities (*arrow*).

C, Postcontrast T1-weighted (400/11/2) coronal image shows no evidence of enhancement of the lesion.

D, This photomicrograph (100 ×) of the patient's lymph node demonstrates follicular hyperplasia and vascular proliferation, which penetrates the germinal center (*large arrow*). There is hyalinization of the central blood vessels (*small arrow*) as well as an onionskin appearance of the mantle zone lymphocytes (*curved arrow* and *star*).

E, Low-power photomicrograph (40 ×) of the lymph node hilum shows central (*curved arrow*) and radial (*arrows*) fibrosis as well as sinus histiocytosis.

Although localized Castleman disease most often occurs as a solitary mass in the mediastinum, it may occur in any area of the body where lymph nodes are found such as the lung, neck, axilla, mesentery, pelvis, and retroperitoneum (2). The MR findings of Castleman disease have been described as nonspecific, low signal on T1-weighted images and high signal on T2-weighted images (12–16). Linear hypointense signals in an arborizing pattern seen within the mass have been attributed to calcification, fibrous septations, or vessels (12). Although Castleman disease has been reported as highly vascular on angiography, other reports have shown no evidence of enhancement on computed tomography scans (13). Our case did not

show enhancement on MR after gadopentetate dimeglumine administration and was not hypervascular at surgery. The internal arborizing pattern seen on MR corresponded to histiocytes and fibrosis within central sinusoids. No calcifications were seen.

We propose that with the presence of central stellate hypointensities in cervical lymph nodes on MR examinations, Castleman disease should be considered.

References

1. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymph-node hyperplasia resembling thymoma. *Cancer* 1956;9: 822–830
2. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer* 1972;29:670–681
3. Denenberg S, Levine PA. Castleman's disease: the lymphoma imposter. *Laryngoscope* 1984;94:601–604
4. Lin JJ, Kirchner FR, Farha GJ. Castleman's disease: surgical management of the head and neck. *J Kans Med Soc* 1978;79(9): 503–506
5. Williams JL, Kaude JV. Sonographic findings in a case of Castleman disease of the neck. *J Ultrasound Med* 1986;5:593–594
6. Kurihara K, Hashimoto N. Extramedullary plasmacytoma associated with a Castleman's lesion of the cervical nodes. *J Oral Pathol* 1983;12:131–138
7. Schroder W, Gnepp DR. Pathologic quiz case 1. *Arch Otolaryngol Head Neck Surg* 1985;111:772–775
8. Anard VK, House JR. Isolated Castleman's disease in the neck. *Ear Nose Throat J* 1989;68:864–869
9. Wolf M, Kessler A, Horowitz A. Benign angiofollicular lymph node hyperplasia (Castleman's disease) presenting as a solitary cervical mass. *J Oral Maxillofac Surg* 1991;49:1129–1136
10. Penfold CN, Cotrell BJ, Talbot R. Neonatal giant lymph node hyperplasia (Castleman's disease) presenting in the head and neck. *Br J Oral Maxillofac Surg* 1991;29:110–111
11. Loftus JM, Jord CN, Hafez GR. Pathologic quiz case 1: Castleman's disease, hyaline vascular type. *Arch Otolaryngol Head Neck Surg* 1991;117:224–226
12. Luburich P, Nicolau C, Ayuso MC, Torra R, Clavers JA. Pelvic Castleman disease: CT and MR appearance. *J Comput Assist Tomogr* 1992;16:657–659
13. de Greer G, Webb WR, Sullitto MC, Golden J. MR characteristics of benign lymph node enlargement in sarcoidosis and Castleman's disease. *Eur J Radiol* 1986;6:145–148
14. Bartkowski DP, Ferrigni RG. Castleman's disease: an unusual retroperitoneal mass. *J Urol* 1988;139:118–120
15. Debatin JF, Spritzer CE, Dunnick NR. Castleman disease of the adrenal gland: MR imaging features. *AJR Am J Roentgenol* 1991; 157:781–783
16. Gianaris PG, Leestma JE, Cerullo LJ, Butler A. Castleman's disease manifesting in the central nervous system: case report with immunological studies. *Neurosurgery* 1989;24:608–612