CASE SNIPPETS

Castlemans disease: retroperitoneal tumor, lichen planus and erythema multiforme

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We report a 32-year-old man with Castlemans disease with retroperitoneal tumor, erosive lichen planus and erythema multiforme, whose skin lesions improved after excision of the retroperitoneal tumor. [Indian J Gastroenterol 1998; 17: 107]

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Retroperitoneal tumor with Castlemans disease is a rare entity. We present such a case with associated skin lesions which improved after excision of the tumor.

A 32-year-old patient presented with painful oral ulcers since 3 months and erosions with itching on the genitalia and extremities since 2 months. He also had odynophagia, remittent fever, continuous dull pain in the left abdomen, and redness of eyes since 2 months. He had lost over 7 kg within 3 months. He had one episode of convulsions 15 days after admission, occasional slurring of speech and tetanic contraction of fingers. There was no history of exposure to sexually transmitted diseases or tuberculosis. There were no other symptoms of alimentary or urinary tract disorder and he was not on any immunosuppressive agents.

On examination, he was aceric with bilateral inguinal, axillary, submandibular and right parotid lymph nodes. There was pterygium formation on the nails, violaceous papules over the extremities with scratch marks, oral and genital erosions, planar ulcers, and blisters with crusting on the palms (suggestive of lichen planus and erythema multiforme). Systemic examination was unremarkable.

Investigations: Blood count, renal function tests, and coagulation profile were normal; ESR was 100 mm (Westergren). Serum bilirubin was normal. AST 63 IU. ALT 52 IU. alkaline phosphatase 1135 IU/L (normal 35-130); serum VDRL was nonreactive. Serum calcium was normal. HIV test was negative. Fine-needle aspiration cytology of the axillary node showed reactive hyperplasia. Biopsy of the skin lesions showed irregular acanthosis, basal cell vacuolar degeneration and lichenoid infiltrate (suggestive of interphase dermatitis resembling lichen planus). Direct immunofluorescence showed fluorescent cytid bodies, a linear epidermal junction and intermittent intercellular cementing substance pattern with IgG, IgM and C3 (suggestive of paraneoplastic pemphigus/lichen planus).

Ultrasoundography showed mild hepatosplenomegaly and a mixed echogenic mass near the splenic flexure of the colon, compressing the left renal pelvis and causing hydronephrosis. CT scan showed a retroperitoneal, contrast-enhancing mass with areas of calcification and hypodensity, compressing the left renal pelvis and causing hydronephrosis. CT scan-guided fine-needle aspiration cytology showed a tumor of spindle cell or round-cell origin. Superior and inferior mesenteric angiography showed no hypervascularity of the tumor. Pre-operative treatment for the erosive lichen planus consisted of proper oral hygiene and prednisolone.

Fig: Interfollicular thick-walled blood vessel undergoing hyalinization (H & E, 100X) (40 mg/day) for 1 week. At surgery a 15 cm x 5 cm x 3 cm retroperitoneal tumor compressing the left ureter and kidney was seen. Para-aortic lymphadenopathy was present. Frozen section of the mass and lymph nodes showed sinus histiocytosis. Excision of the tumor and left kidney was done. Para-aortic nodes were sampled. The postoperative recovery was uneventful. The oro-cutaneous lesions decreased significantly within 15 days.

Gross examination showed a multinodular tumor weighing 400 grams surrounded by dense fibro-fatty tissue, and a hydro nephrotic kidney. The tumor was firm with focally distributed areas of calcification. Microscopy revealed large lymphoid follicles with ill-defined germinal centers. The lymphoid cells were arranged concentrically around them. The interfollicular and medullary vessels were thick-walled, showing hyalinization (Fig). The small vessels in the germinal centers also showed hyalinization.

Castlemans disease is a rare benign form of lymphoma, which can present as localized or generalized varieties.1 The former presents with symptoms of compression in the mediastinum or abdomen. The generalized form presents with fever, anaemia, weight loss, lymphadenopathy, altered liver function, renal disease, convulsions, or cutaneous lesions, paraneoplastic pemphigus, lichen planus, erythema multiforme and keratoconjunctivitis.1 It may be associated with systemic lupus erythematosus, Sjogren’s syndrome, cardiomyopathy, rheumatoid arthritis or plasma celloma.1

Remission occurs with excision of the localized tumor,2 as in our patient. These lesions can progress to malignant lymphoma.2 Chemotherapy, radiotherapy and autologous bone marrow transplant have been used to treat the diffuse variety, though the prognosis is poor.2

References

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