

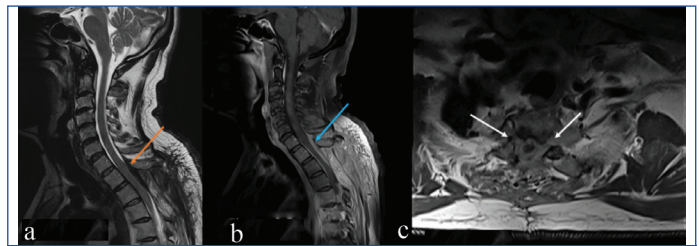
IgG4 Related Disease Presenting as a Lung Mass: Imaging Findings

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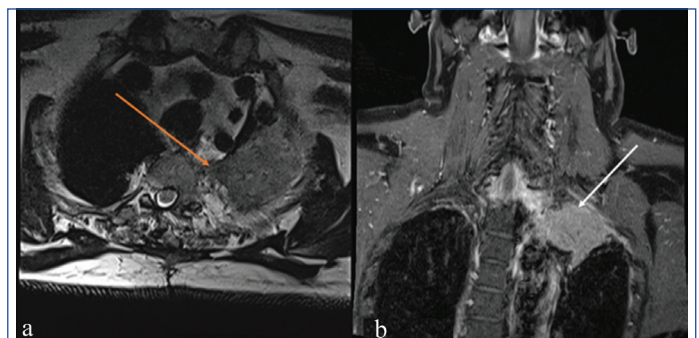
Keywords: C-reactive protein, Cord compression, Cortical erosion, Extradural lesion, Lower limb weakness

A 60-year-old male patient presented with complaints of left upper limb weakness and sensory loss, mild left lower limb weakness for five days, accompanied by difficulty in passing urine for three days. He was previously admitted to another institute for similar complaints, where he was diagnosed to have D2-D3 spinal extradural lesion with cord compression, for which he underwent D2-D3 laminectomy with excision of the lesion. At that time the patient recovered well and the histopathology report showed evidence of IgG4 related disease. The patient's general condition was fair and vitals were normal with blood pressure of 110/70 mmHg and pulse rate of 85 beats per minute in present admission. Neurological examination revealed significant motor weakness (power 3/5) and mild sensory deficits in the left upper limb. There was also mild weakness in bilateral lower limbs on examination (power 4/5). Laboratory investigations revealed anaemia (haemoglobin: 10.8 g/dL; reference range for males: 13-17 g/dL) and a markedly elevated C-reactive protein level (>130 mg/L; reference value: <10 mg/L), suggesting an active inflammatory process. He was subjected to a chest X-ray, plain CT of the thorax, and contrast-enhanced MRI of the dorsal spine. Chest X-ray showed an ill-defined radio-opaque mass in the left upper zone [Table/Fig-1a]. Plain CT chest showed subtle cortical erosion involving the posterior aspect of the left first rib [Table/Fig-1b]. Plain and contrast-enhanced MRI of the dorsal spine revealed a T2 hypointense thickening of the dura of the spinal cord from C7 to D2 vertebral bodies. There was associated extradural intense enhancing soft tissue at that level, extending into bilateral D1-D2 neural foramina [Table/Fig-2a-c]. The mass on the left-side was extending to involve the upper lobe of the left lung with formation of a relatively well-defined mass measuring 5.7×4.8 cm in the left upper lobe with contrast enhancement [Table/Fig-3a,b]. CT guided biopsy from the left upper lobe lung mass was done, and specimen was sent for Histopathological Examination (HPE). Histopathology of the lung biopsy specimen from the left upper lobe confirmed interstitial lung disease characterised by plasma cell-rich inflammation, fibrosis, and obliterative vasculitis, which were consistent with IgG4-related lung disease. Serum IgG4 levels were elevated at 189 mg/dL (reference range: <135-140 mg/dL). The patient was treated with intravenous methylprednisolone 1 g once daily for five days, resulting in symptomatic improvement,

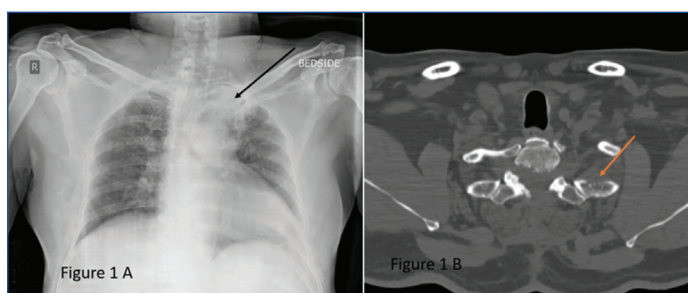
and was subsequently discharged. Presently, the patient has partially recovered and can do his day-to-day activities.



[Table/Fig-2a-c]: a) MRI of the dorsal spine revealed a T2 hypointense thickening of the dura of spinal cord from C7 to D2 vertebral bodies (Orange arrow); b,c) There was associated extradural intense enhancing soft-tissue at that level (blue arrow), extending into bilateral D1-D2 neural foramina (white arrows).



[Table/Fig-3a,b]: a) Axial; and b) coronal image of MRI chest showed a well-defined mass in the left upper lobe with contrast enhancement (Orange arrow in axial image and white arrow in coronal image).



[Table/Fig-1a,b]: a) Chest X-ray shows ill-defined radio-opaque mass in the left upper zone (black arrow); b) Plain CT chest showed subtle cortical erosion involving the posterior aspect of 1st rib (Orange arrow).

IgG4 related disease is an inflammatory condition characterised by infiltration of IgG4 positive plasma cells and fibrosis in various organs. It can affect different organs and mimics malignancy [1]. Commonly involved organs include the salivary glands (chronic sclerosing sialadenitis), thyroid (Riedel thyroiditis), mediastinum (fibrosing mediastinitis), pancreas (autoimmune pancreatitis), orbit (orbital pseudotumour), biliary tree (sclerosing cholangitis), and retroperitoneum (retroperitoneal fibrosis) [1]. In the present case, it involved the cervicodorsal spine and adjacent lung. The clinical symptoms of a patient depend upon the organ involved. In the present case, he had a history of left upper limb weakness and sensory loss, and mild left lower limb weakness. Salmaggi A et al., reported a case of IgG4RD presenting as spinal compression similar to the present case [2]. Fu L et al., reported CT findings of paravertebral tissue involvement adjacent to thoracic vertebrae involved by IgG4-related disease, similar to the present case [3]. The pathogenesis remains unclear, with the hypothesis of an autoimmune disease being the most widely accepted [4]. An increase in serum IgG4 levels has a role in the diagnosis of IgG4-RD. But it has some limitations, as IgG4 levels are not always elevated, and an increase in IgG4 levels is also associated with other conditions such as bronchiectasis, asthma, sarcoidosis, and adenocarcinoma of the pancreas. So, increase in serum IgG4 levels is considered to have high sensitivity and positive predictive value (>90%) but with low specificity and

negative predictive value [4]. Patients with infiltration of more than one organ or aggressive conditions show elevation of acute phase markers, such as C-Reactive Protein (CRP) and Erythrocyte Sedimentation Rate (ESR) which constitute the main markers of disease activity or relapse, like the present case [5]. Diagnostic confirmation of IgG4-RD by biopsy is strongly recommended for the exclusion of malignancies and other IgG4-RD mimics. Lee LIT et al., reported a case of isolated pulmonary IgG4-related disease mimicking lung malignancy [6]. In the present case lung lesion was simulated like a malignancy with subtle rib erosion. But a lung biopsy confirmed the diagnosis of IgG4-related disease. Stamatopoulos A et al., reported a case of IgG4-related lung disease extending to the thoracic vertebra [7]. In the present case, IgG4-related spinal lesion was seen extending into the adjacent lung.

Radiologist should consider IgG4-RD as a differential diagnosis in patients with an epidural mass. Prompt treatment with high dose corticosteroid is necessary to avoid long-term neurological complications.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Feb 07, 2026
- Manual Googling: Feb 19, 2026
- iThenticate Software: Feb 21, 2026 (1%)

ETYMOLOGY: Author Origin

EMENDATIONS: 5

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Dec 24, 2025**

Date of Peer Review: **Feb 09, 2026**

Date of Acceptance: **Feb 23, 2026**

Date of Publishing: **Jul 01, 2026**