

A Rare Case of Mediastinal Gray Zone Lymphoma Presenting With Tracheoesophageal Fistula

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Introduction

- Mediastinal gray-zone lymphoma (MGZL) is an extremely rare variant of lymphoma with characteristics intermediate between Nodular sclerosis Hodgkin lymphoma (NSHL) and primary mediastinal B-cell lymphoma (PMBL).
- Mediastinal lymphomas rarely present with tracheoesophageal fistula (TEF), and to date, there are no reported cases of MGZL presenting with TEF.
- Here we report a case of an elderly female presenting with an invasive mediastinal mass forming a TEF diagnosed as MGZL.

Case presentation

- A 78-year-old female presented with a 1-week history of cough, expectoration, fevers, chills, shortness of breath, odynophagia, scanty hemoptysis, and weight loss.
- At presentation, she was hypertensive, tachycardic, afebrile, tachypneic, and hypoxic with room air saturation of 88%.
- On examination, bilateral crackles were present on lung auscultation.
- CT of the chest with contrast showed innumerable bilateral pulmonary nodules, a large conglomerate mediastinal adenopathy (measuring 5.5 x 4.0 x 9.2 cm) encasing the trachea and esophagus (Fig. 1), multiple enlarged right paratracheal lymph nodes, and bilateral pleural effusion.
- Upper GI endoscopy showed a large, ulcerating, infiltrating mass invading and fistulizing into the middle third of the esophagus (Fig. 3). A fully covered esophageal stent measuring 23 mm x 12.5 cm was placed across the tumor (Fig. 4), and a gastrostomy tube was also inserted.
- Biopsy and HPE revealed large pleomorphic lymphoid cells with a spectrum of cytologic features resembling those of multinucleated Reed-Sternberg cells, lacunar cells, centroblasts, or immunoblasts (Fig. 5).
- Immunohistochemistry (IHC) showed positivity for CD20, CD79a, MUM1, PAX5, P53, CD30, BCL6, and CD45, and negativity for CD15, ALK, CD10, EBV (LMP), MART1, and pan-keratin, consistent with MGZL (Fig. 6-9).
- Due to her age and frailty, she was started on R-mini-CHOP (Rituximab and Reduced-Dose CHOP) and was discharged after the first cycle of chemotherapy.

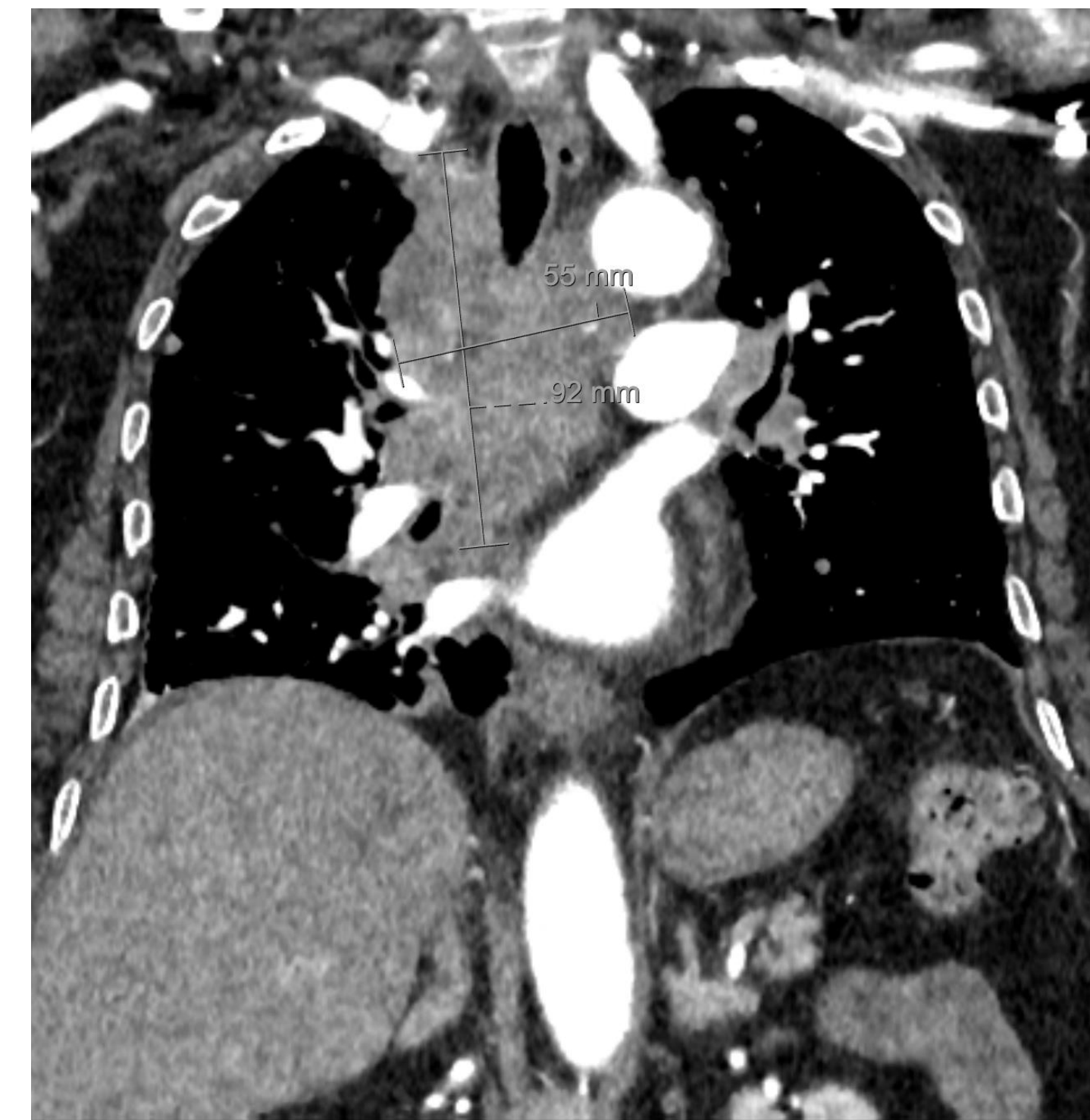


Figure 1 - CT of the chest with contrast on coronal section shows a large conglomerate mediastinal adenopathy measuring 5.5 x 4.0 x 9.2 cm encasing the trachea and esophagus

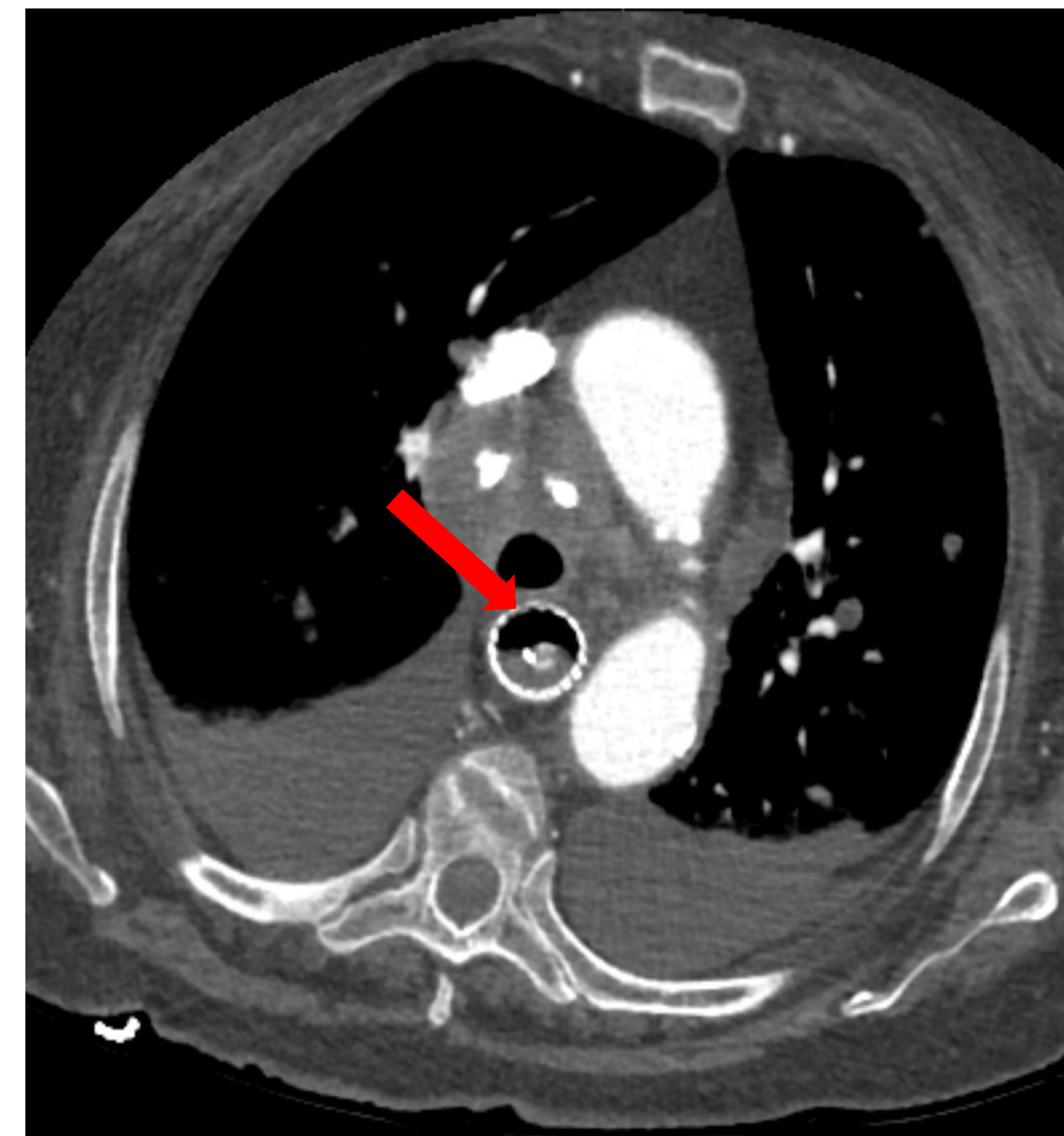


Figure 2 - CT of the chest with contrast on axial section shows the esophageal stent placed across the tumor (red arrow)

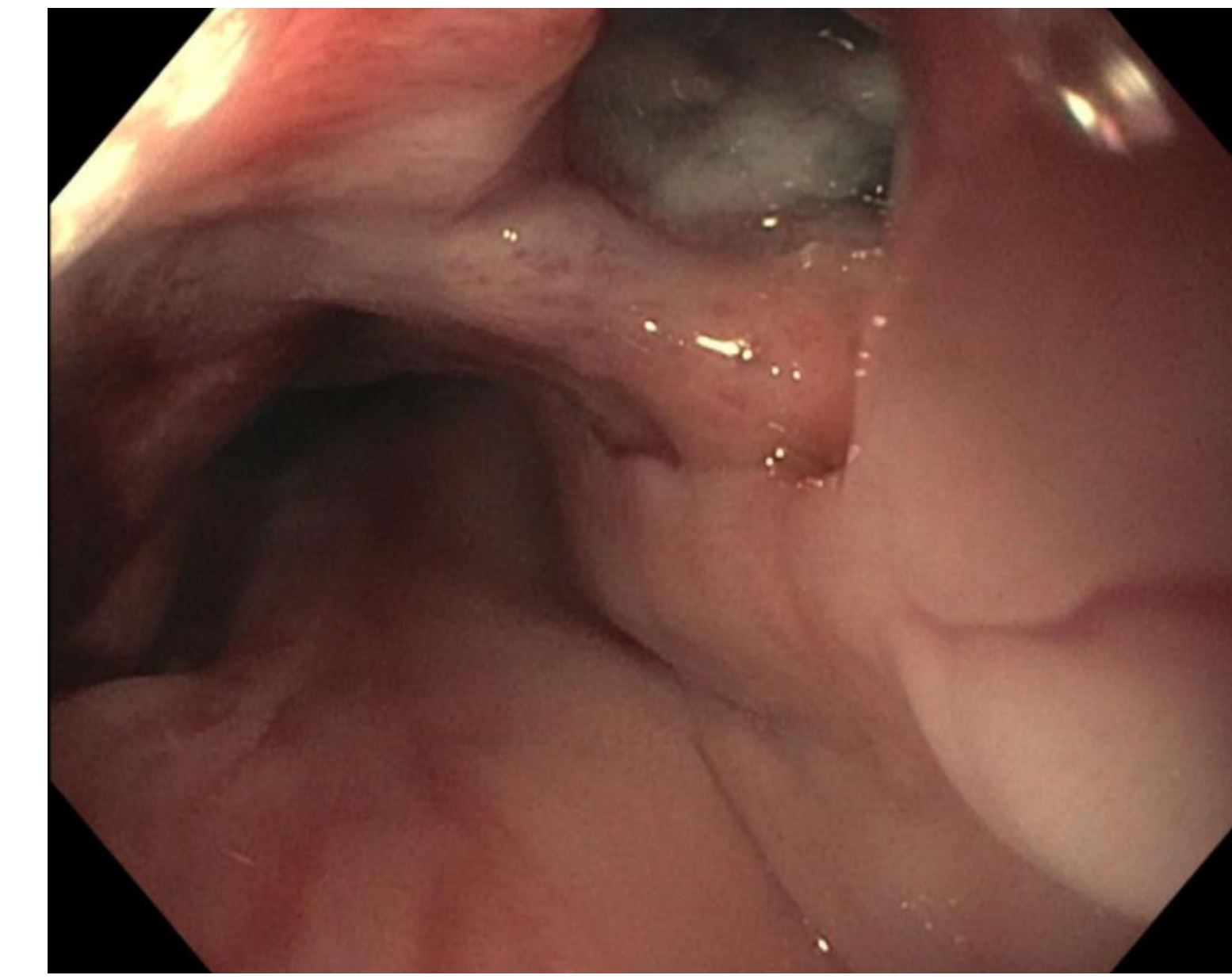


Figure 3 - a large, ulcerating, infiltrating mass invading and fistulizing into the middle third of the esophagus

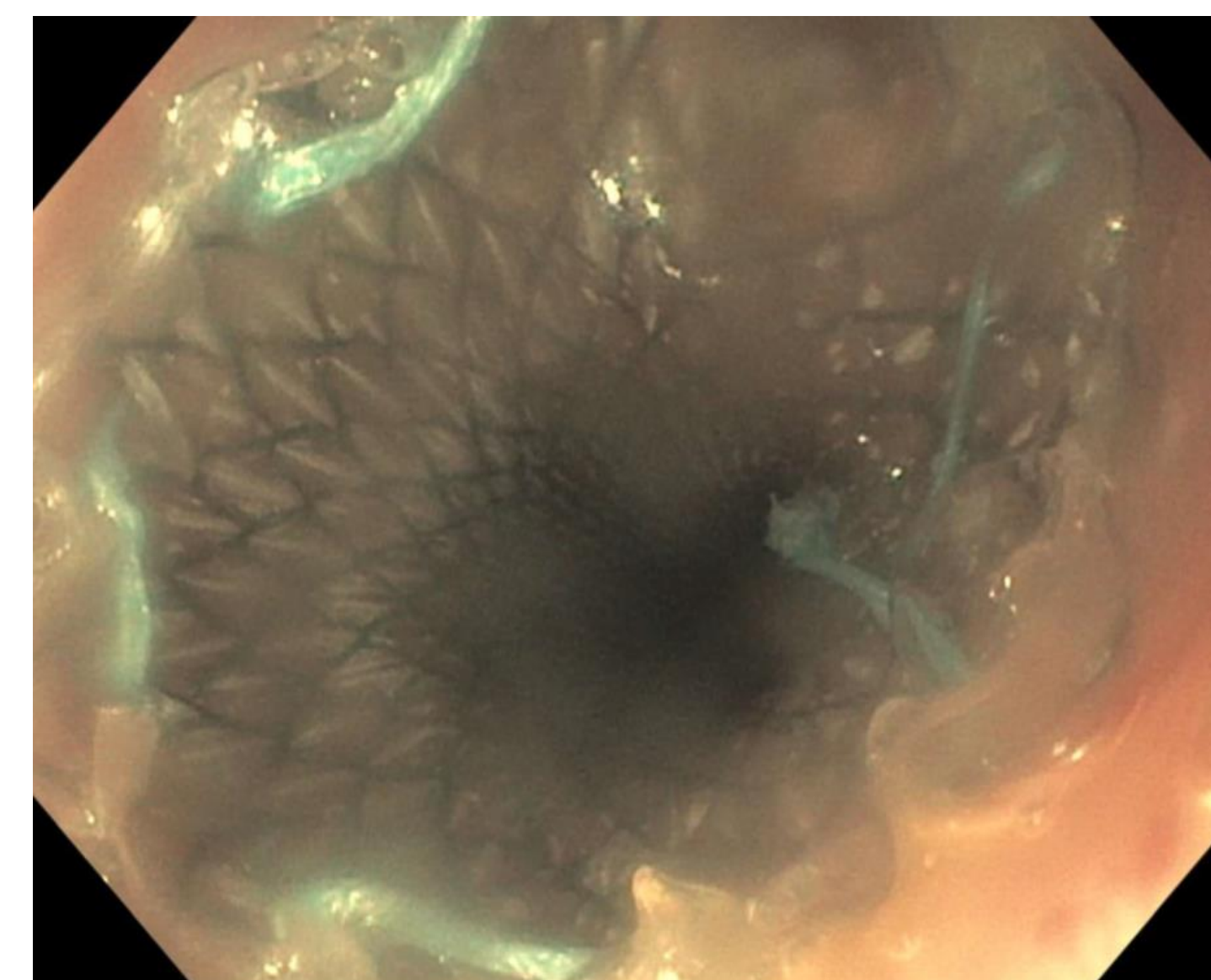


Figure 4 - A fully covered esophageal stent measuring 23 mm x 12.5 cm was placed across the tumor

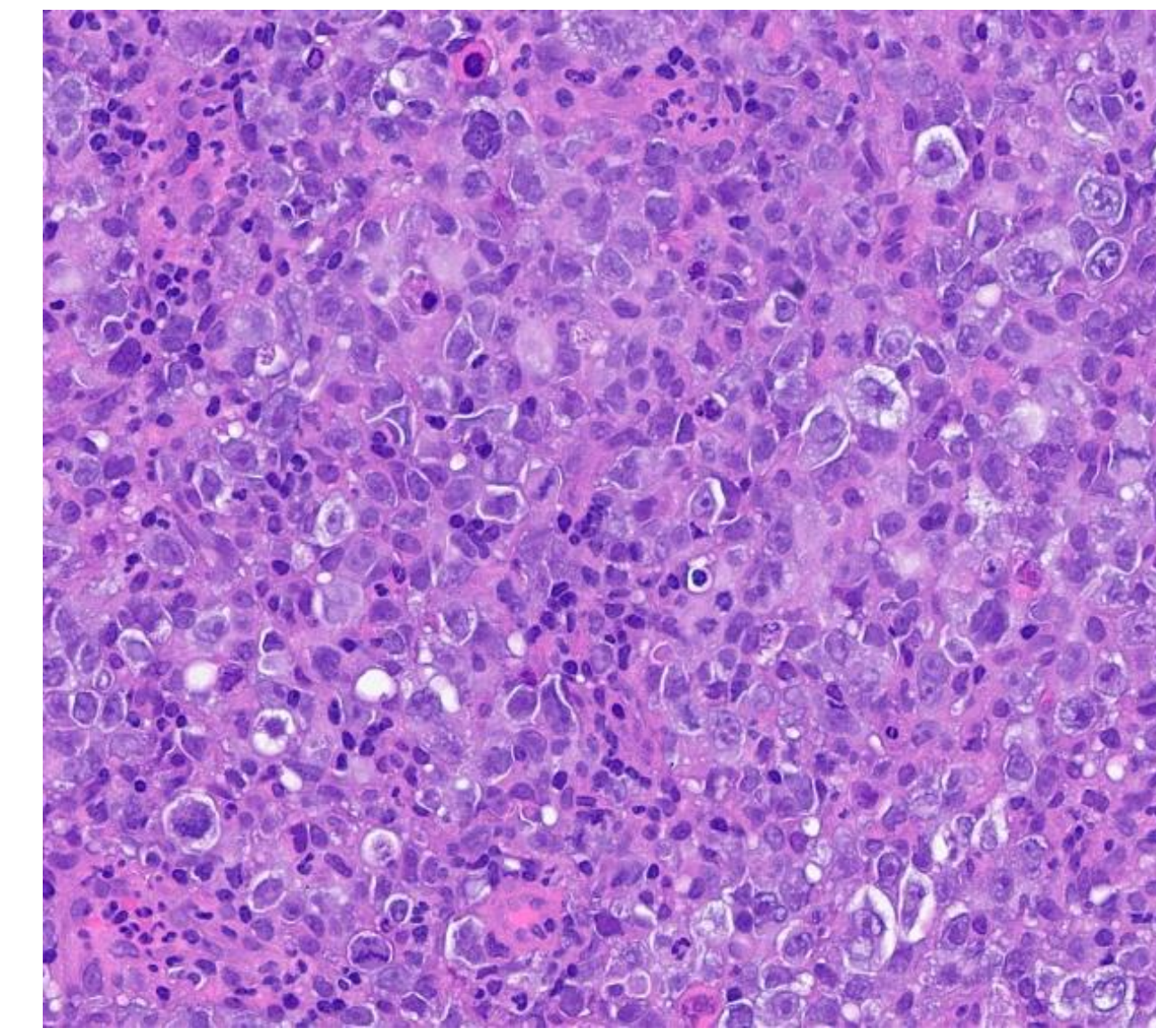


Figure 5 – HPE (200X) shows large pleomorphic lymphoid cells with a spectrum of cytologic features resembling multinucleated Reed-Sternberg cells, lacunar cells, centroblasts, or immunoblasts

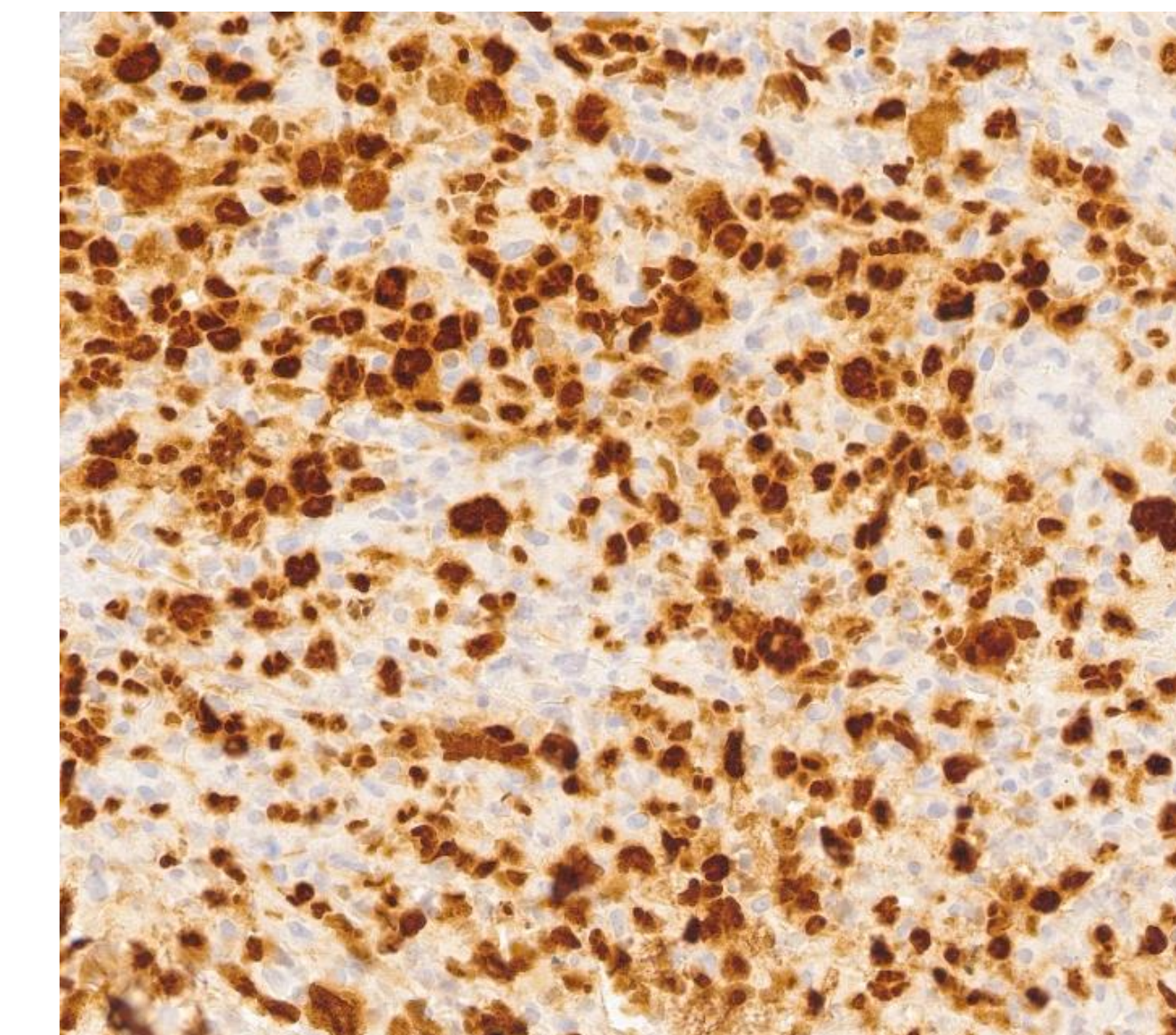


Figure 6 – IHC (200X) - the atypical lymphoid cells are positive for MUM1

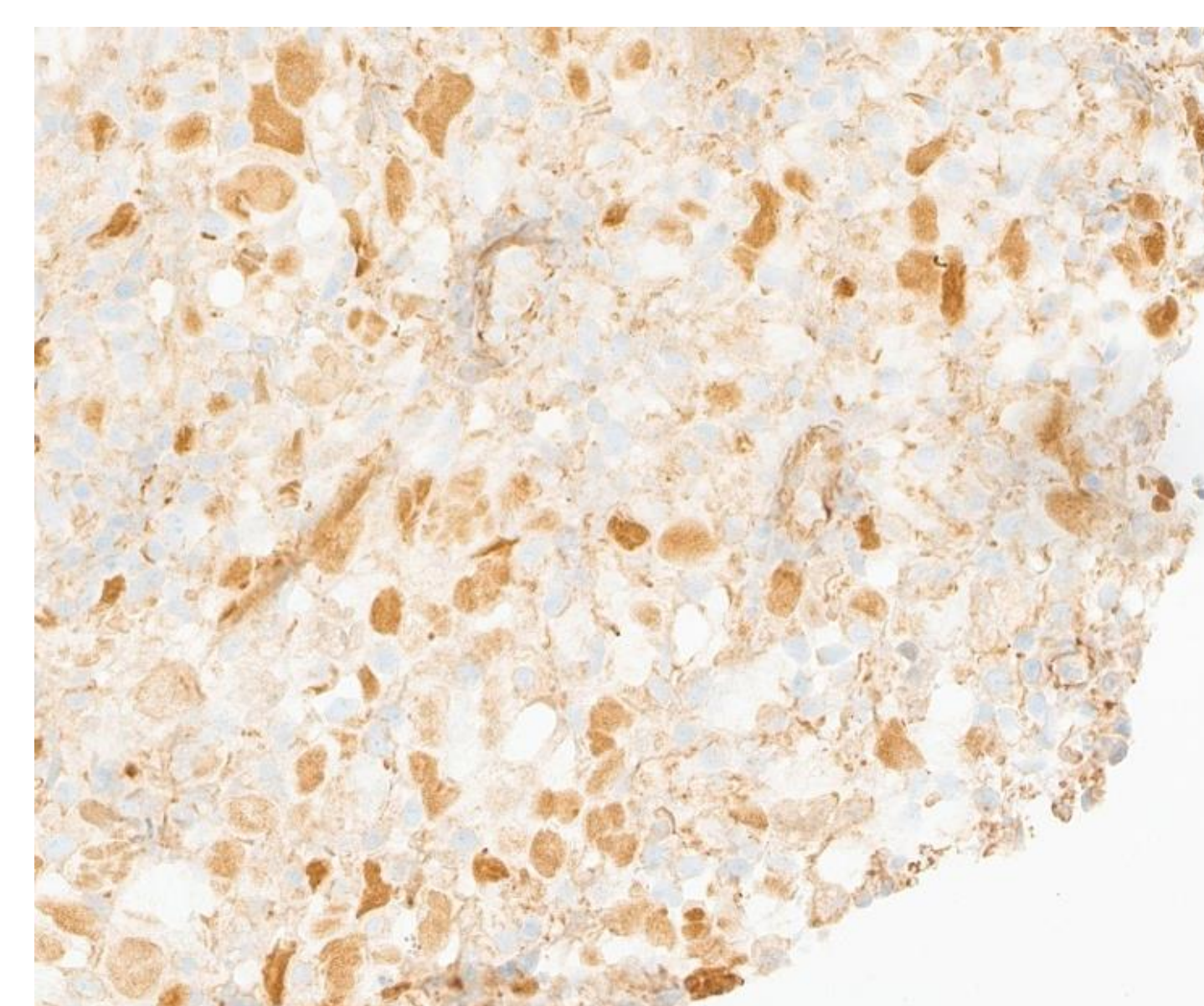


Figure 7 – IHC (200X) - positive for PAX-5

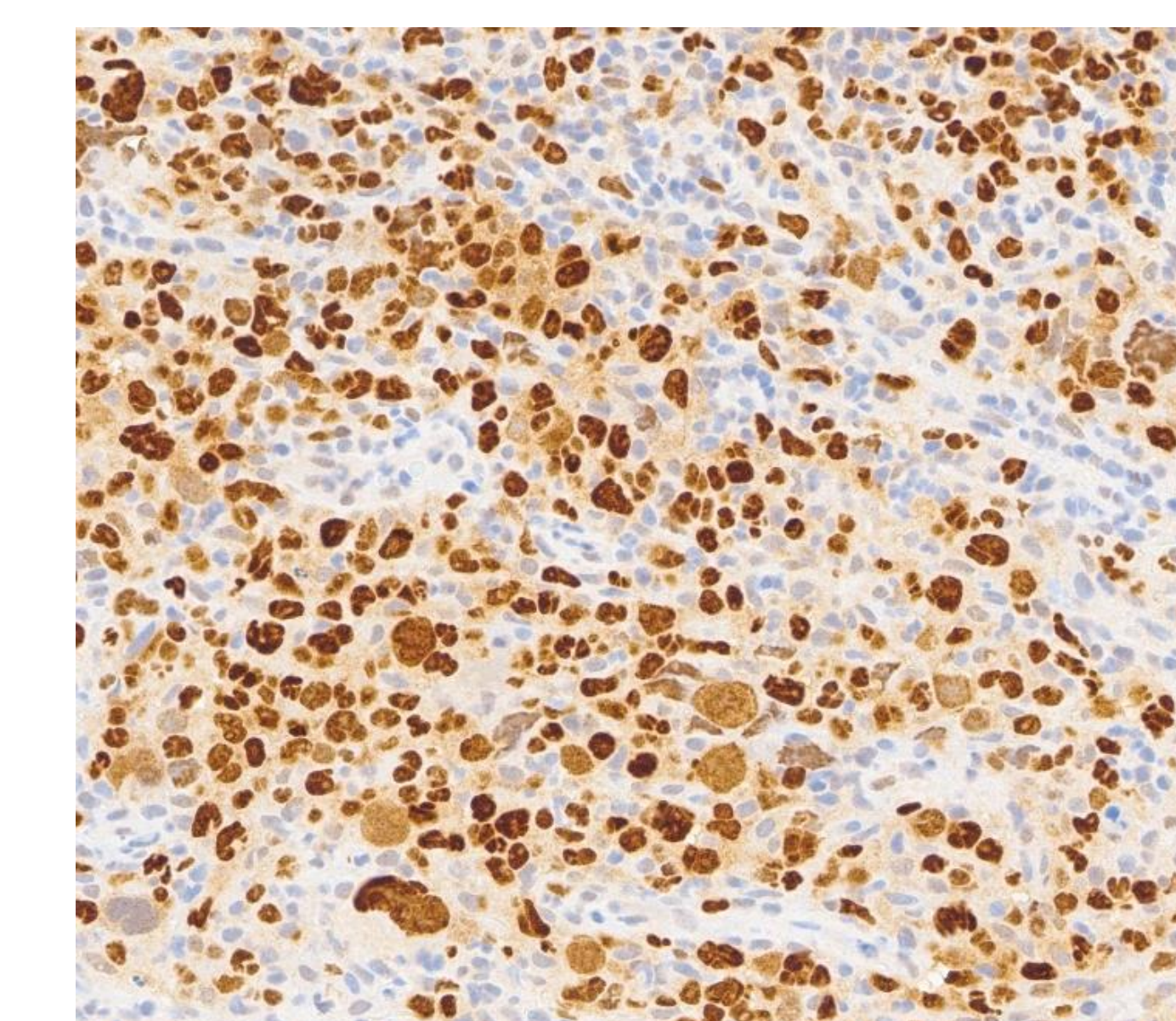


Figure 8 – IHC (200X) - positive for p53

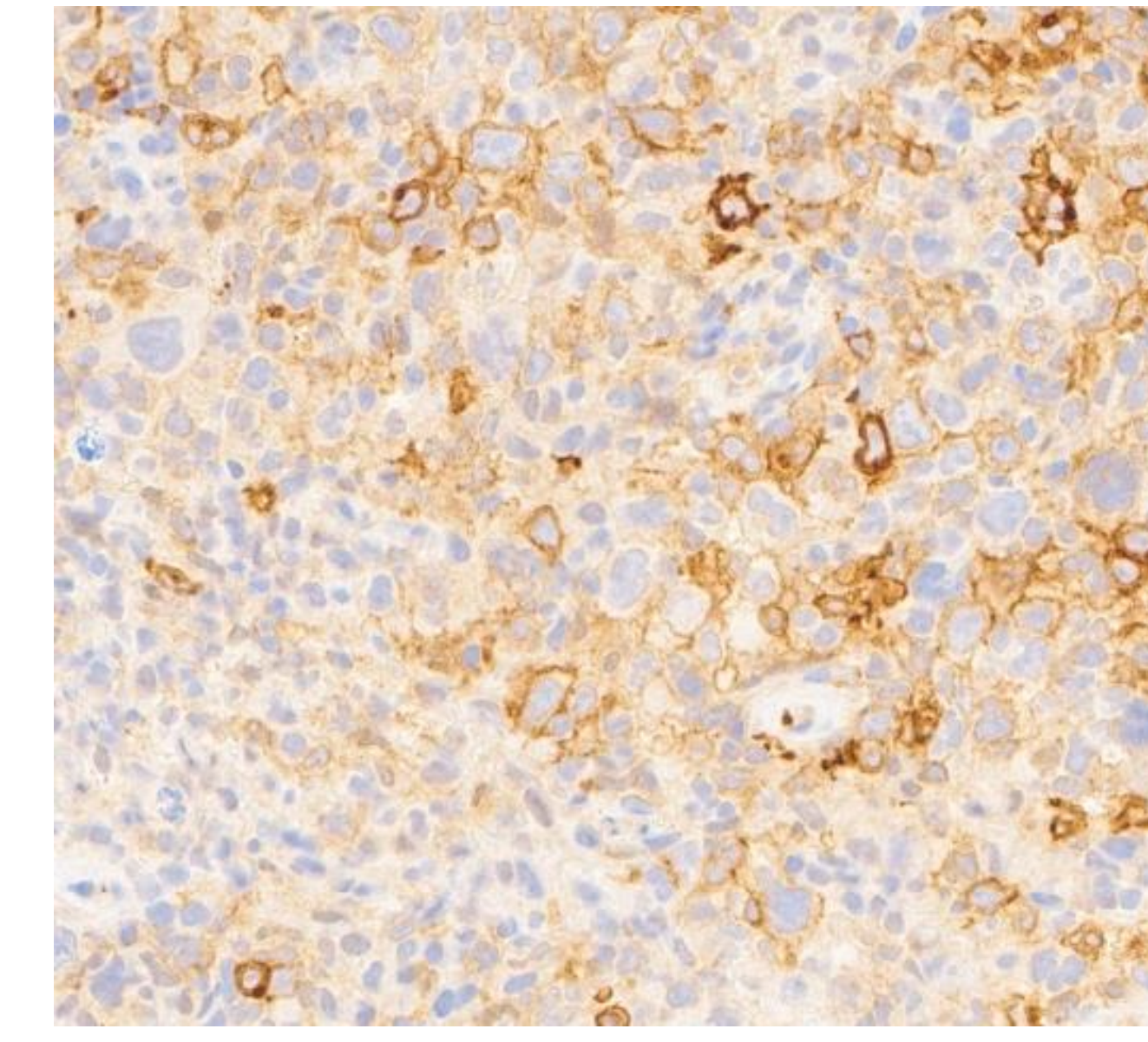


Figure 9 – IHC (200X) - A significant proportion expresses CD30

Discussion

- MGZL is most frequent in young adult males (median age 33 years). (1) Our patient, being 78-year-old female at the time of diagnosis, was unique.
- Dose-adjusted etoposide, doxorubicin, and cyclophosphamide with vincristine, prednisone, and rituximab (DA-EPOCH-R) is an effective treatment for MGZL, with event-free survival and overall survival being 62% and 74%, respectively. (2) Due to the patient's condition, we started R-mini-CHOP. (2)
- Brentuximab with nivolumab has shown promise in relapsed or refractory MGZL with potential bridge to stem cell transplant. (3)
- Malignant TEF is common with esophageal and lung cancers but is rarely seen in mediastinal lymphoma.
- It most commonly involves the upper two-thirds of the esophagus, as in our case.
- Treatment needs to be individualized but is usually managed with an esophageal and/or an airway stent, and gastrostomy to prevent possible aspiration pneumonia and mediastinitis.
- The majority (67%) of the fistulas may close after treatment with chemoradiation. (4)
- TEF related to lymphoma has a better prognosis than those caused by esophageal or lung cancers.

Conclusion

- TEF in mediastinal lymphoma requires early identification and prompt treatment with a tracheal or esophageal stent.
- A gastrostomy further helps to prevent aspiration pneumonia and mediastinitis.
- Prompt initiation of chemotherapy often closes the TEF.
- Due to its rarity, evidence for the treatment of MGZL is limited, but DA-EPOCH-R provides the best outcomes, and further research is required to explore other novel therapies.

References

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