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CASE REPORT
Cardiac imaging

Case report: multimodality imaging to diagnose cardiac diffuse large B-cell lymphoma

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Background Primary cardiac lymphoma (PCL) is rare and can present with a wide variety of clinical symptoms, frequently leading to a delay in diagnosis.

Case summary We report a case of a PCL in an 81-year-old man. Cardiac magnetic resonance imaging showed multiple masses in the right atrium and a mass in the right ventricular outflow tract extending to the pulmonary artery. Biopsy revealed a diffuse large B-cell lymphoma. The patient also had metastases to the liver and lung on the positron emission tomography–computed tomography (PET–CT) scan. He was treated with R-CHOP chemotherapy, with complete remission documented PET–CT scans.

Conclusion Although most patients with PCL die before chemotherapy can be initiated, a timely diagnosis can result in a favourable outcome.

Keywords Cardiac lymphoma • Cardiac MRI • Echocardiography • Primary cardiac tumour

Learning points

- Primary cardiac tumours are rare, can present with atypical symptoms and sometimes presenting a diagnostic challenge.
- Primary cardiac lymphomas (PCLs) arise from the right side of the heart in the majority of cases.
- Early diagnosis of PCL is important, potentially leading to early treatment and a better prognosis.
- PCL may be cured by chemotherapy depending on the type and promptness of diagnosis.

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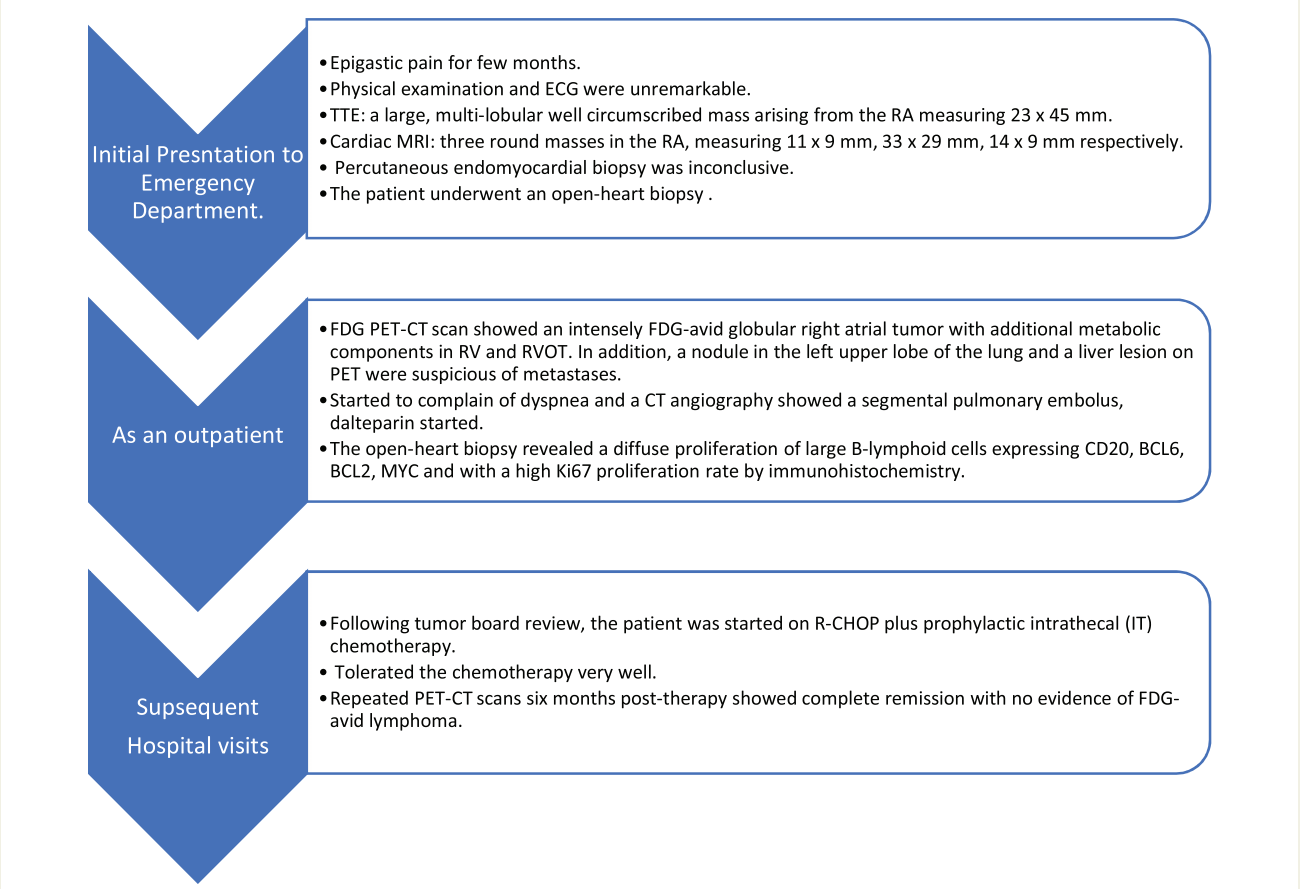
Introduction

Primary cardiac lymphoma (PCL) is a rare disease and sometimes presents with non-specific clinical symptoms that make the diagnosis challenging.[1] We are presenting an interesting case that required multimodality imaging and procedures to establish the diagnosis.

Timeline



Case presentation

The 81-year-old Caucasian male with no significant past medical history presented to our emergency department with epigastric pain for a few months. Physical examination and electrocardiogram were unremarkable. Transthoracic echocardiography showed normal left ventricle (LV) dimensions and function (estimated ejection fraction 60–65%), a mildly dilated right ventricle (RV) with low-normal systolic function, and mild mitral regurgitation. There was a large, multi-lobular well-circumscribed mass arising from the right atrium (RA) wall laterally, just below the tricuspid annulus, measuring 23 mm × 45 mm (*Figure 1*, *Video 1*). Cardiac magnetic resonance imaging (MRI), showed normal LV and RV function and three round masses in the RA, attached to its wall, measuring 11 mm × 9 mm, 33 mm × 29 mm, and 14 mm × 9 mm, respectively (*Figure 2A*). There was also a round, highly mobile mass on a narrow stalk in the RV outflow tract (RVOT) below the pulmonary valve, attached to the posterior RVOT wall, measuring 23 mm × 17 mm (*Figure 2B*). Additionally, there was a large, intrapericardial mass anterior to the pulmonary artery and the aorta, which appeared to be connected to the mass inside the RVOT. These masses were all heterogeneous on T1- and T2-weighted MRI imaging compared to the myocardium, with enhancement on perfusion scan. Collectively, these findings were highly suggestive of malignancy, although no apparent primary malignancy was identified on history or physical examination. After discussion, the Heart team decided to perform percutaneous endomyocardial biopsy which was inconclusive, hence the patient underwent an open heart biopsy that revealed a diffuse proliferation of large B-lymphoid cells expressing CD20, BCL6,

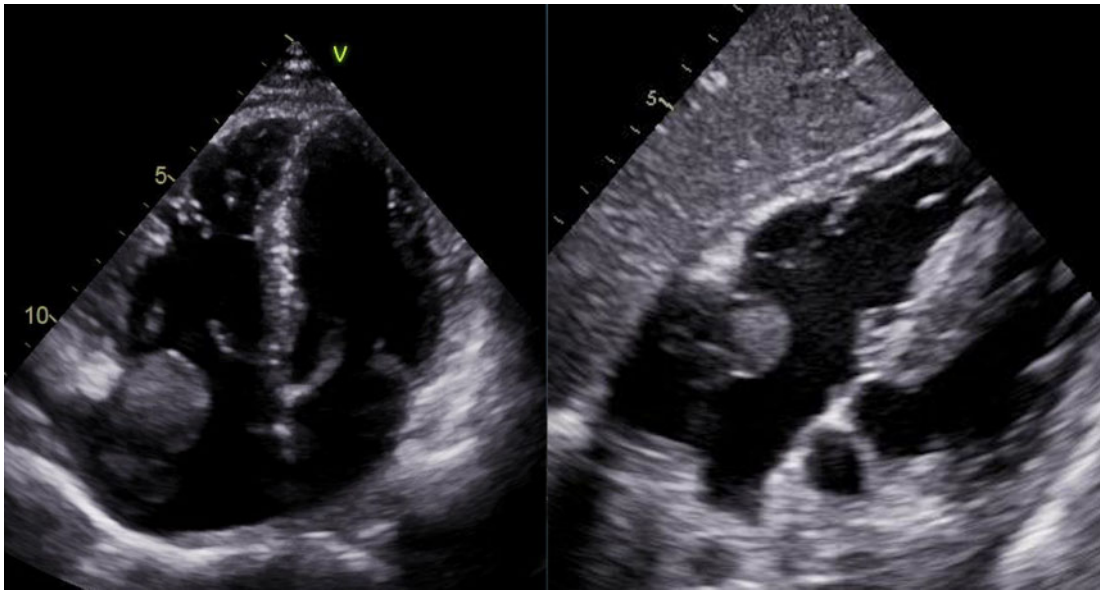


Figure 1 Transthoracic echocardiogram: apical and subcostal four chambers view showing the mass in the right atrium measuring 23 ham× 45 mm.

BCL2, MYC and with a high Ki67 proliferation rate by immunohistochemistry (*Figure 3*), and showing rearrangement of *MYC*, not *BCL2* or *BCL6* by FISH, consistent with diffuse large B-cell lymphoma with *MYC* rearrangement. A whole body ^{18}F fluorodeoxyglucose positron emission tomography–computed tomography (FDG PET–CT) scan showed an intensely FDG-avid globular right atrial tumour with additional metabolic components in RV and RVOT. In addition, a nodule in the left upper lobe of the lung and a liver lesion on PET were suspicious of metastases (*Figure 4*, left). While the patient was waiting for the result of the biopsy, he started to complain of dyspnoea and a CT angiography showed a segmental pulmonary embolus for which he was started on dalteparin. Following tumour board review, the patient was started on R-CHOP plus prophylactic intrathecal chemotherapy. He tolerated the chemotherapy very well. Repeated PET–CT scans (*Figure 4*, right) 6 months post-therapy showed complete remission with no evidence of FDG-avid lymphoma.

Discussion

Primary cardiac lymphomas accounts for 1% of primary cardiac tumours.[1] In the majority of reported cases, PCLs arise from the right side of the heart.[1,2] Patients usually present with non-specific symptoms, thus a challenging diagnosis delaying early treatment.[1–3] Symptoms vary according to the site of involvement and include atrioventricular block (first to third degree), refractory heart failure, and pulmonary embolism.[2–4] Our patient had no specific cardiac symptoms. Although a tissue diagnosis was required, the multi-imaging modality contributed to his early diagnosis and treatment. An open heart procedure was necessary since a percutaneous endomyocardial biopsy failed to confirm the diagnosis. The prognosis of PCLs is generally poor with a median survival of 7 months after diagnosis.[2] However, with a timely diagnosis, patients respond well to chemotherapy as did our patient, who received chemotherapy 2 months from initial presentation, achieving complete remission.

Conclusion

Primary cardiac lymphoma is rare. The diagnosis can be challenging because of a variable and non-specific clinical presentation. Most patients die before chemotherapy can be initiated, but will usually respond well to chemotherapy following a timely diagnosis. In our patient, complete remission was observed on PET–CT scans post-chemotherapy.

Strength and limitations

The strength part of our case is the multimodality approach used to diagnose the case; including echocardiography, cardiac MRI, FDG PET–CT, and the tissue biopsy.

Lead author biography

Yahya E. Alansari is currently a cardiology fellow at McGill University, Montreal, Canada. He is board certified in internal medicine in USA, Canada, and UK (ABIM, FRCPC, MRCP). He did internal medicine residency at the University of Miami, Florida and then moved to Montreal to complete his cardiology training. He is interested in Interventional Cardiology and he published papers and presented abstracts in main cardiology conferences in USA and Canada.

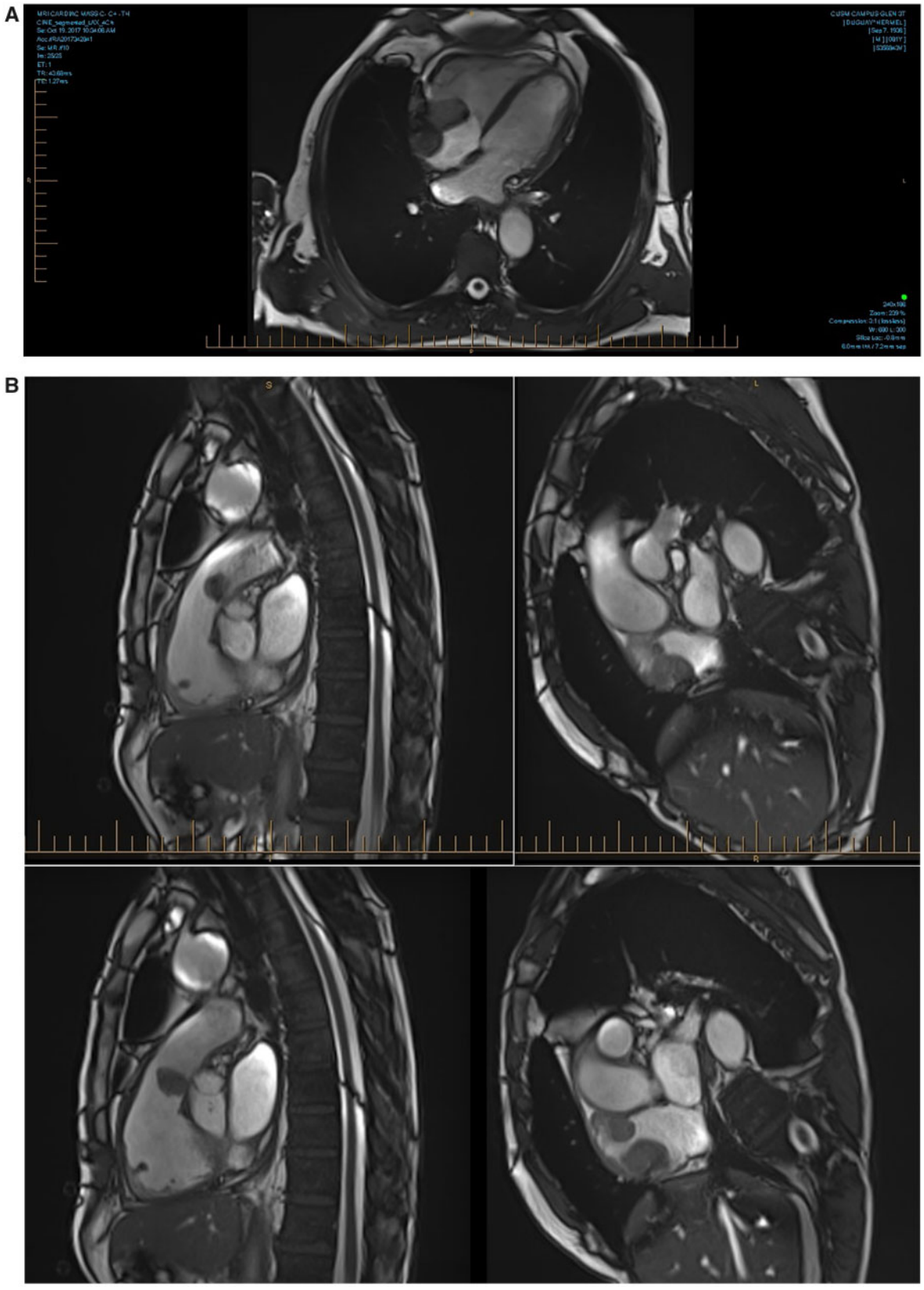


Figure 2 Cardiac magnetic resonance imaging: (A) steady-state free precession of four chambers view showing three round isolated masses in the right atrium, all attached to its wall, measuring 11 mm × 9 mm, 33 mm × 29 mm, and 14 mm × 9 mm, respectively. (B) Different views of steady-state free precession showing the right atrium masses and the right ventricular outflow tract mass right ventricle outflow tract.

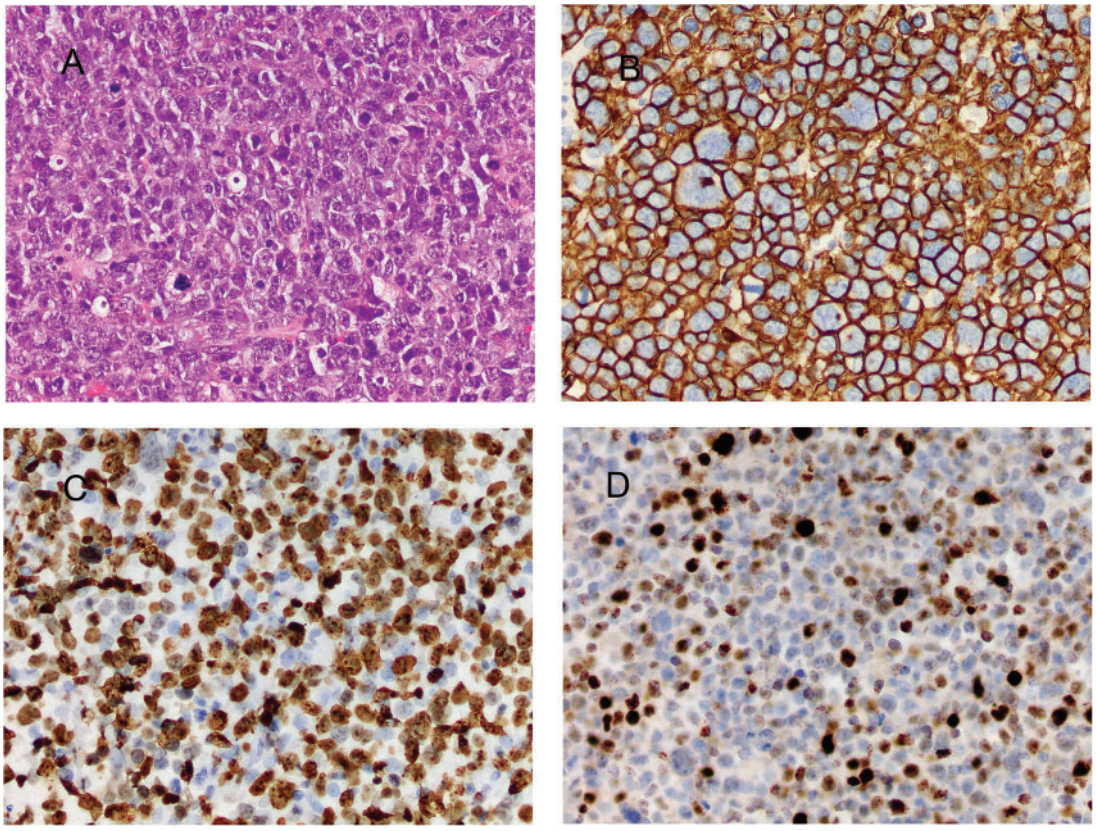


Figure 3 Light microscopy of the patient's diffuse large B-cell lymphoma. (*A*) H&E-stained section shows large lymphoid cells with pleomorphic nuclei, variably prominent nucleoli, and scant to moderate amount of cytoplasm. (*B*) Immunostain for CD20 outlines the membranes of the large B cells. (*C*) Immunostain for Ki67 showing a proliferation rate of 80–90% of neoplastic lymphoid cells. (*D*) Immunostain for MYC, positive in 50–60% of neoplastic cells. All magnifications ×200.

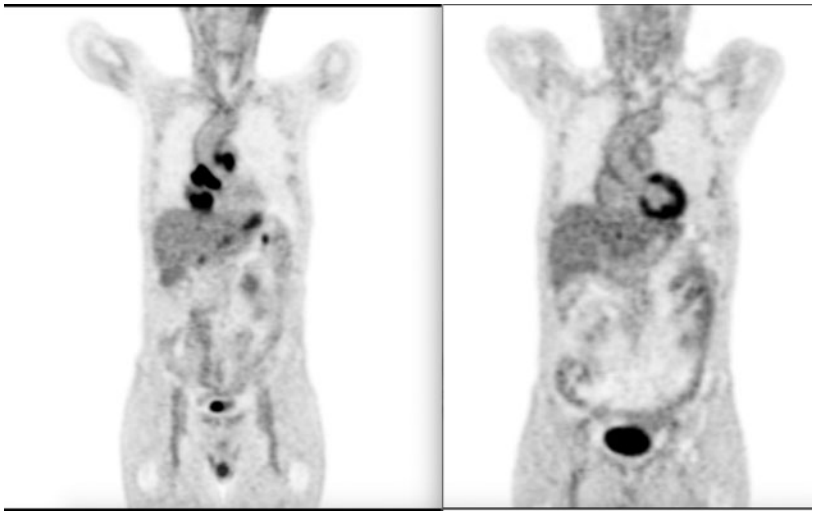
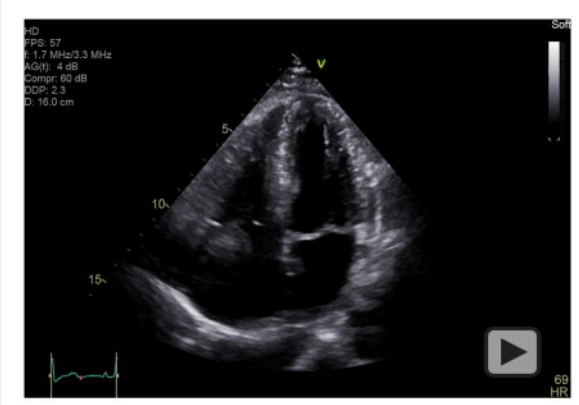


Figure 4 ^{18}F fluorodeoxyglucose positron emission tomography whole body. (Left) Pre-treatment with chemotherapy showed an intensely ^{18}F fluorodeoxyglucose-avid globular right atrial tumour with additional metabolic components in right ventricle and right ventricle outflow tract. In addition, a nodule in the left upper lobe of the lung and a liver lesion on positron emission tomography were suspicious of metastases. (Right) Post-chemotherapy, showed no lymphoma.



Video 1 Transthoracic echocardiogram short clip showing the mass in the right atrium.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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