

Sarcomas: rare primary malignant tumors of the thorax

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ABSTRACT

In this article, it is aimed to review the radiological signs of unusual primary malignant tumors of the thorax. Radiological studies of 11 patients with histologic diagnosis of thoracic sarcomas were interpreted retrospectively. Tumors originated from the chest wall (n=3), mediastinum (n=4), and pulmonary parenchyma (n=4). Histopathologic diagnoses were fibrosarcoma (n=1), alveolar rhabdomyosarcoma (n=1), malignant hemangiopericytoma (n=1), malignant fibrous histiocytoma (n=2), pulmonary vein leiomyosarcoma (n=1), pulmonary artery sarcoma (n=2), pleuropulmonary blastoma (n=1), and chondrosarcoma (n=2). In order to evaluate thoracic sarcomas, cross-sectional methods such as CT and MRI can be useful in demonstrating the origin of the mass, relationship with and involvement of adjacent structures. They present as masses and, unfortunately, radiological findings are not sufficient for specific diagnosis.

Key words: • sarcoma • lung • thorax

Sarcomas are rare tumors of the thorax. They can originate from the lung parenchyma, mediastinum, pleura, or the chest wall. Angiosarcoma, leiomyosarcoma, rhabdomyosarcoma, and mesothelioma (sarcomatoid variant) are the most commonly encountered histopathological types. The diagnosis is established after making the differential diagnosis for histopathologically sarcoma-like malignancies (sarcomatoid carcinomas) and metastatic disease. Radiologically, these lesions usually appear as large heterogeneous masses. However, their radiological appearance can range from an intrabronchial or intravascular mass to a solitary pulmonary nodule (1).

Herein, we review the radiological findings of the cases diagnosed with malignant primary mesenchymal tumors of the thorax in our hospital between 1996 and 2002, and compare them with the data available in the literature.

Malignant fibrous histiocytoma

Malignant fibrous histiocytoma is the most frequently observed soft tissue sarcoma in adults. It generally develops after irradiation therapy. It is rarely localized within the thorax (1). Frequently, it originates from the muscles of the chest wall and seldom from the lungs, mediastinum, or pleura (1-3). It is usually a disease of advanced age and is seen during the sixth and seventh decades of life. It does not have any gender preference. Most of the cases are asymptomatic (1). They appear as well-defined, regular or lobulated soft tissue masses and do not have any characteristic radiological features (Figures 1 and 2). They enhance heterogeneously with contrast material and rarely contain calcifications (1-3) (Figure 1).

Chondrosarcoma

Chondrosarcoma is the most commonly observed primary tumor of the chest wall. It is more common in males and in the age group of 30-60. It typically originates from the sternum or costochondral regions in the anterior chest wall (1, 4, 5). It can, on occasion, originate from the lung parenchyma or bronchi. It is the most frequently encountered malignant tumor of the sternum (6). These tumors appear as large expanding masses containing chondroid type calcifications accompanied by a significant soft tissue component, causing destruction of the bone (Figures 3 and 4).

Rhabdomyosarcoma

Rhabdomyosarcoma is a childhood tumor having bimodal age distribution. In adults, it is seen after the age of 50 and is more frequent in males (1). It can originate from any part of the thorax and the presence of striated muscle tissue is not a prerequisite for its development (1, 7).

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Received 29 May 2003; revision requested 19 September 2003; revision received 24 May 2004; accepted 01 June 2004.

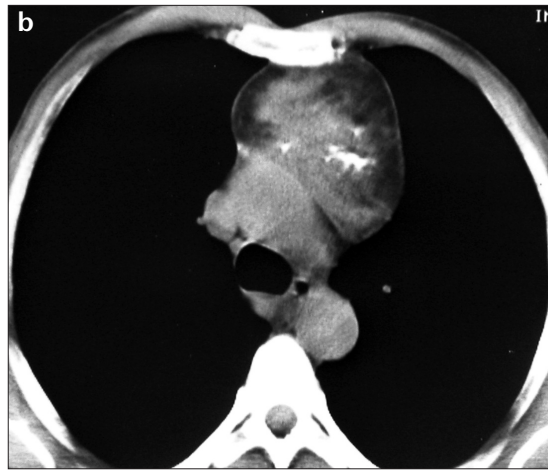
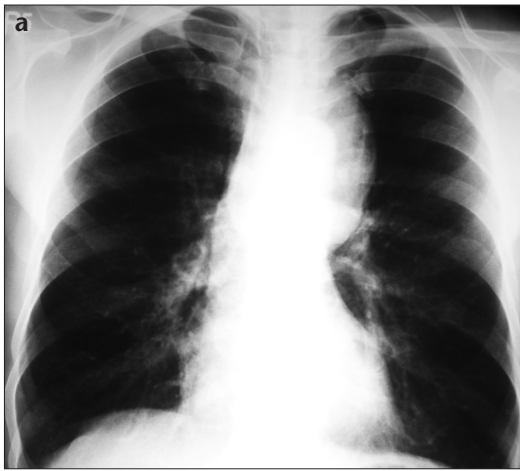


Figure 1. a, b. Malignant fibrous histiocytoma. A 53-year-old male with dyspnea presented with an anterior mediastinal mass with calcifications. Posteroanterior chest radiograph (a) and CT section (b). There are cases reported in the literature as anterior mediastinal masses (3).

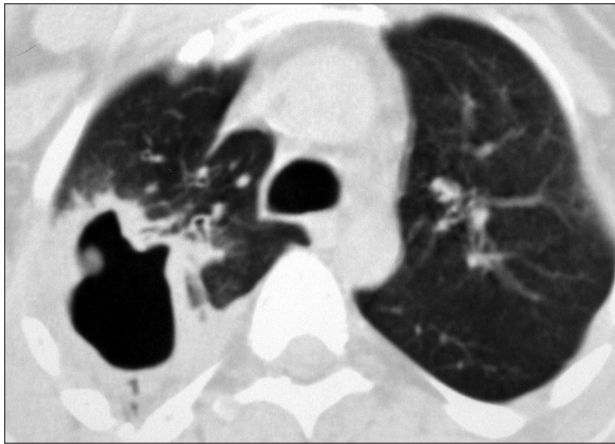


Figure 2. Malignant fibrous histiocytoma. A 39-year-old male with hemoptysis. CT section shows a cavity with thick and irregular walls in the right upper lobe.

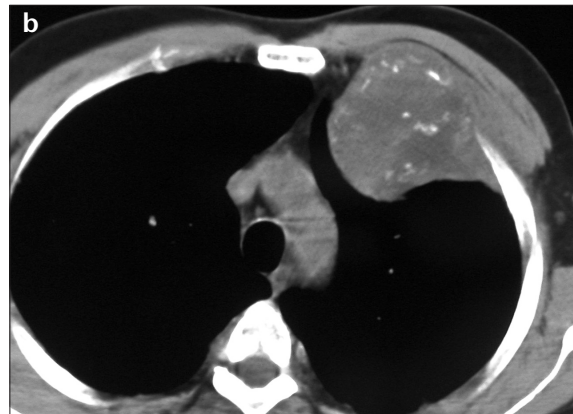
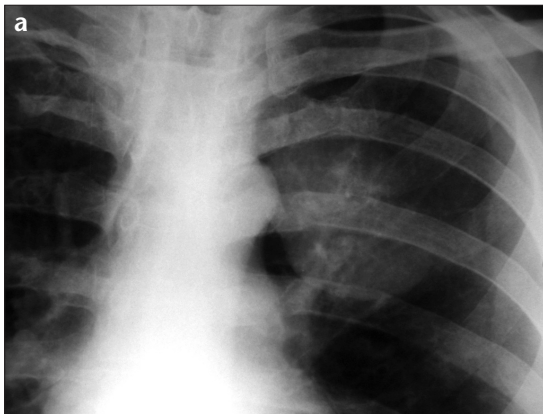


Figure 3. a, b. Chondrosarcoma. A 45-year-old male with chest pain. Posteroanterior chest radiograph (a) and CT section (b) show an expansile mass with relatively regular margins containing calcifications originating from the left second costochondral region.

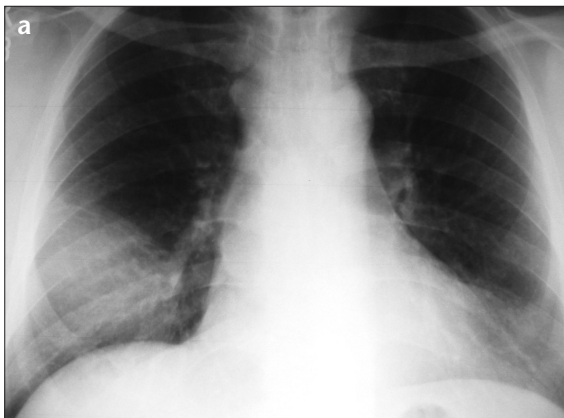


Figure 4. a, b. Chondrosarcoma. A 51-year-old female with dyspnea was found to have an expansile rib mass with a calcified matrix. Posteroanterior chest radiograph (a) and CT section (b).

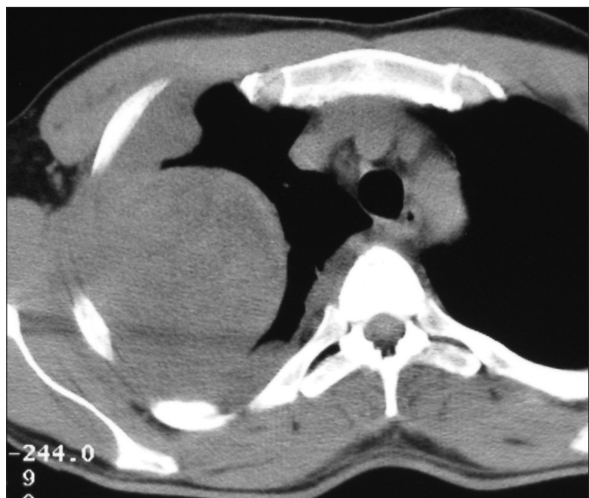


Figure 5. Rhabdomyosarcoma (alveolar type). A 35-year-old male with cough and dyspnea. CT section. Right hemithorax contains a lobulated mass extending throughout the pleural space.

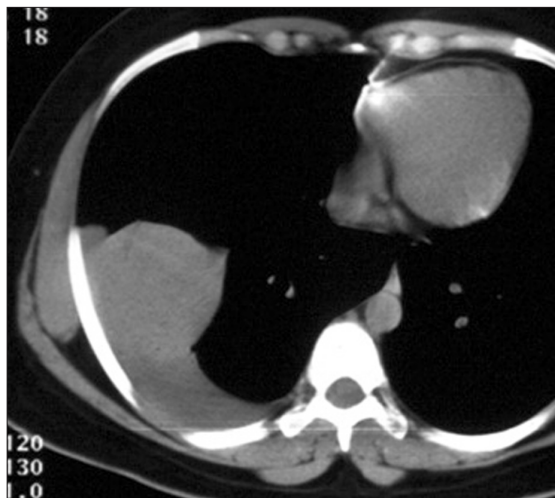


Figure 6. Fibrosarcoma. A 27-year-old female with dyspnea. CT section. A mass was observed in the right lung extending to the fissure.

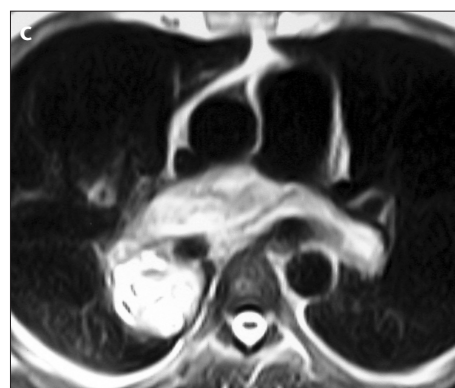
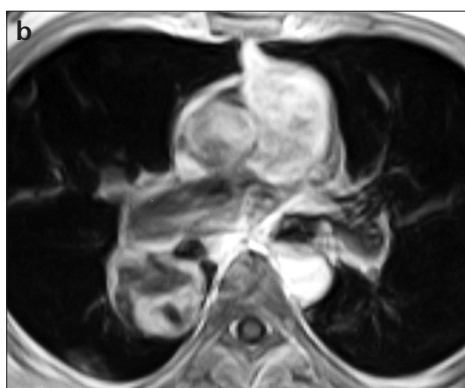
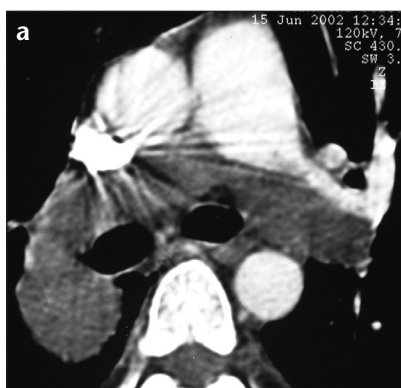


Figure 7. a-c. Sarcoma of the pulmonary artery. CT examination (a) of a 52-year-old male with the complaint of dyspnea demonstrates a hypodense mass filling both main pulmonary arteries and mimicking massive pulmonary embolism. MR imaging (b, c) shows that the mass enhances homogeneously with contrast material and contains areas of necrosis.

However, it most frequently originates from the chest wall or diaphragm in the adult age group (1). It is the most commonly observed sarcoma of the heart in children (8). Unlike Ewing sarcoma and primitive neuroectodermal tumors, chest wall rhabdomyosarcomas generally do not metastasize to the ribs (9). Prognosis depends on the histopathological subtype. Alveolar type has a more unfavorable prognosis compared to the embryonic and pleomorphic subtypes. Thoracic location is also an adverse prognostic factor (1, 7).

Since these tumors can remain clinically silent, they may have already reached large dimensions by the time of diagnosis. This is especially true for those masses originating from the chest wall (Figure 5) (1, 7). There may be necrosis and cystic areas within the mass. The tumors can extend into or

invade the neighboring vessels and bronchi. Those originating from the heart present as hypodense masses within the chambers of the heart and become symptomatic at an earlier stage (1).

Fibrosarcoma

Fibrosarcoma develops from the connective tissue found in the structures of the thorax. It is most commonly observed in children and young adults. In adults, it generally originates from the chest wall and lungs. In children, it develops as an intraluminal mass within the main or lobar bronchi and can cause obstructions. As a result, nearly all of these lesions are symptomatic (1, 10). It is also seen frequently in the left atrium and can lead to heart failure (8).

Radiological findings differ depending on the site of involvement. Intra-

bronchial tumors are associated with findings of atelectasia, whereas those originating from the chest wall form large masses. Fibrosarcomas originating from the lungs are similar to other solid lesions in appearance (Figure 6). Foci of calcification or ossification may be observed on CT (1).

Pulmonary artery sarcomas

Pulmonary artery sarcomas generally originate from the main or proximal pulmonary arteries. They are seen around the age of 50 and have no gender preference. Their clinical symptoms and radiological findings are similar to those of pulmonary emboli, which consequently cause delays in diagnosis (1, 5).

They enlarge throughout the lumen within the vessel (5). Radiologically, they appear as polypoid masses within the lumen (Figures 7 and 8). They can

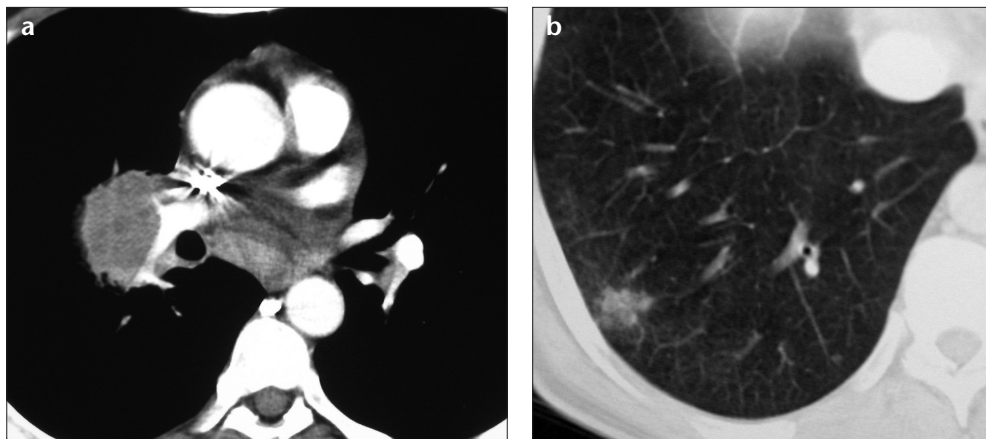


Figure 8. a, b. Pulmonary artery leiomyosarcoma. A 52-year-old male with cough and weight loss. CT examination (**a, b**) demonstrates a mass within the right pulmonary artery (**a**). Metastatic nodules are seen (**b**) in the right lower lobe. At surgery, several metastatic nodules were identified in the lung and pleura.

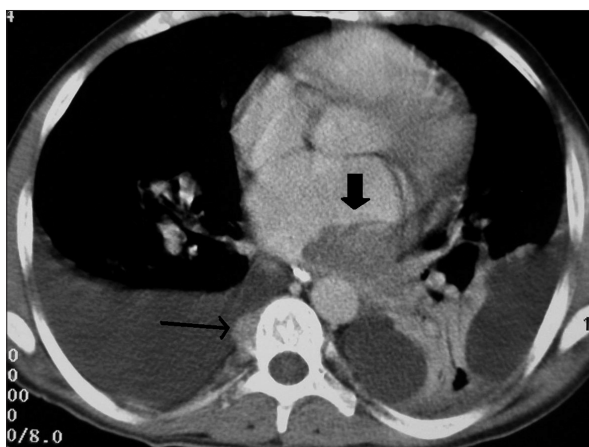


Figure 9. Pulmonary vein and cardiac leiomyosarcoma. A 49-year-old thalassemic male suspected to have pulmonary embolism. CT shows a mass extending from the left inferior pulmonary vein to the atrium (**bold arrow**). Note extramedullary hematopoietic focus in the right paravertebral region (**thin arrow**).

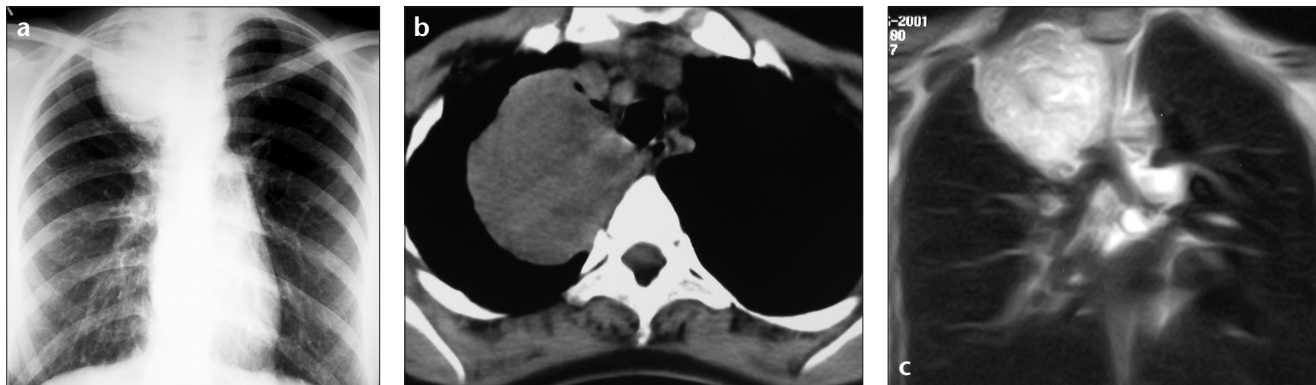


Figure 10. a-c. Pleuropulmonary blastoma. A 23-year-old female with dyspnea and cough. Posteroanterior chest radiograph (**a**), CT section (**b**), and coronal MR image (**c**). Note the lobulated solid mass (type III) in the right apex with extrapulmonary localization.

invade the surrounding structures by extension from the vessel wall. Contrast enhancement in MR imaging may aid in the differential diagnosis from emboli (Figure 7). They have poor prognosis with a life expectancy of 6-12 months (1).

Cardiac leiomyosarcomas that have a preference for the left atrium are easily differentiated from angiosarcomas that favor the right atrium (Figure 9). Cardiac leiomyosarcomas have a poor prognosis. In the radiological examina-

tion of leiomyosarcomas, ECG-triggered MR imaging is helpful for evaluating the relationship with neighboring vital structures (1, 4, 8).

Pleuropulmonary blastoma

Pleuropulmonary blastoma (PPB) is a rare tumor of childhood. It can originate from the lungs as well as from tissues other than the lungs. It is claimed to be of mesodermal origin. There are three histopathological subtypes: cystic (type I), mixed solid and cystic

(type II), and solid (type III). Type I has the best prognosis. Types II and III have poor prognosis with 5-year survival rates as low as 35%. The cystic type can easily be mistaken for other benign cysts. It is reported that there is a relationship to other cystic diseases of the lung such as congenital cystic adenomatoid malformation, extralobar sequestration, bronchogenic cyst and PPB (11, 12).

Radiologically, they appear as masses with peripheral localization, but large

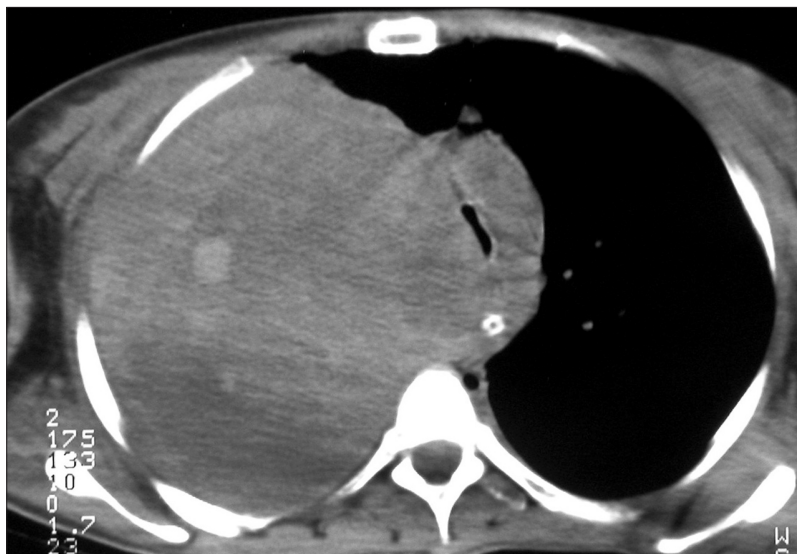


Figure 11. Hemangiopericytoma. A 28-year-old female with chest pain and dyspnea. CT shows a mass filling the right hemithorax and compressing the mediastinal structures.

tumors can involve a whole lobe (Figure 10). They may be adherent to the visceral pleura. The right lung is frequently involved and they can spread to hilar or mediastinal lymph nodes (11). They can metastasize to the same lung, central nervous system, and skeletal system (11, 12).

Malignant hemangiopericytoma

Hemangiopericytoma is thought to originate from the pericytes of the small vessels (4, 13). It is seen in adults and does not have any gender preference. Generally, these tumors are in the form of solitary, well-defined masses without any relationship to the airways or vessels. They can reach large dimensions (Figure 11). Symptoms are not specific (5, 13). They can cause paraneoplastic findings like hypoglycemia (13, 14).

Radiologically, they present as well-defined peripheral or central masses. Necrosis or calcifications may be found. Despite having a vascular origin, they have limited blood supply. They generally have a benign character; however, on rare occasions, they can invade the mediastinum or the chest wall (5).

Conclusion

When a mass is identified within the thorax, the potential diagnosis

to be first considered is the common pulmonary malignancies such as adenocarcinoma, squamous cell carcinoma, and undifferentiated large cell or small cell carcinomas. However, although they are rare, sarcomas originating from the lung parenchyma, mediastinum, chest wall, or pleura should be considered in the differential diagnosis (1, 13). Different histopathological types of sarcomas cannot usually be differentiated on radiological grounds, though the site of localization of the mass and clinical findings might aid in the diagnosis. Ewing sarcoma, primitive neuroectodermal tumor, chondrosarcoma, malignant fibrous histiocytoma, osteosarcoma, synovial sarcoma, and fibrosarcoma generally originate from the chest wall. For example, in the case of a large mass of costal origin that is accompanied by fever in a child, Ewing sarcoma should be the primary consideration, while a rib mass with a calcified matrix may represent a chondrosarcoma or an osteosarcoma. A mass of pulmonary artery origin should lead to the diagnosis of a primary pulmonary artery sarcoma like leiomyosarcoma (1).

In the radiological evaluation of the rarely encountered tumors of the thorax, cross-sectional imaging techniques are helpful in defining the origin of

the mass, identifying the relationships with the surrounding structures, and its spread; yet, radiological findings are variable and are not usually lesion-specific.

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