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Case report

Primary malignant melanoma of the lung in oculocutaneous albino patient

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Abstract

Oculocutaneous albinism is a rare autosomal recessive disorder characterized by general depigmentation, nystagmus, photophobia, and decreased visual acuity. Malignant melanoma is extremely rare in patients with albinism. We present a 41-year-old albino male patient, who was admitted with a suspected bronchogenic carcinoma. He underwent a pulmonary resection and the diagnosis was primary malign melanoma of the lung. The patient died of tumor recurrence in the postoperative 46th month. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Albinism; Malign melanoma; Lung

Oculocutaneous albinism is a rare autosomal recessive disorder occurring as a result of lack of tyrosinase enzyme [1,2]. Its incidence in Caucasians is 1:39,000 [1]. The entity is characterized by albinism, pink-white skin, white hair, grey–blue eyes color, nystagmus, and photophobia. Patients with albinism lack protection against sun-light, thus UV induced skin cancers are frequently encountered [2]. In contrast, malignant melanoma occurrence is very rare [1,3,4].

Malignant melanoma consists 1% of all malignant tumors and 3% of malignant tumors of the skin [5]. Extracutaneous malignant melanoma is a rare entity that can originate from squamous mucous membranes, eyes, esophagus, and leptomeninges [2].

1. Case report

A 41-year-old albino male patient was admitted to our clinic with complaints of cough, hemoptysis, and dyspnea.

In the physical examination, albinism, nystagmus, and a 1×1.5 cm sized hyperemic, crusted lesion at the extensor face of the left forearm were observed. Routine laboratory studies and biochemical tests were normal. Chest X-ray showed a mass at the para-hiler field and an approximately 6×12 cm sized mass located in the left lower lobe near the left hilus (Fig. 1) and a nodule, 1 cm in diameter, in the superior segment of the right lower lobe were determined at the thorax CT scan. The nodule in the right lung was considered as benign because of its being calcified. A cranial CT scan, an abdominal ultrasonography, and a whole body bone scintigraphy revealed no abnormalities. A polypoidal tumor arising from the left lower lobe bronchus was seen via bronchoscopy. Biopsy showed a non-small cell carcinoma. Excisional biopsy was performed for the lesion at the left forearm and the histopathological assessment resulted as a basal cell carcinoma. Considering histopathological results and the fact that basal cell carcinoma metastasis is a rare event, presence of two separate primary malignancies was accepted.

The patient underwent left lower lobectomy with the diagnosis of a non-small cell bronchogenic carcinoma. Two masses in the left lower lobe were detected; a 7×7 cm sized one at the apical segment of the left lower lobe and a 2×3 cm sized one at the anterolateral segment of the left lower lobe, and left lower lobectomy with complete hilar and mediastinal lymph dissection was performed.

Histopathological examination of the specimen revealed that tumor cells had oval or spindle nuclei prominent one or more eosinophilic macro-nuclei, and eosinophilic, spindle or polygonal cytoplasm some also containing brown–black pigments. There were typical and atypical many mitotic figures. Another nodular lesion macroscopically and histopathologically resembling the bronchial one, was detected in the parenchyma beneath the visceral pleura.

The lymph nodes resected from the hilar region were

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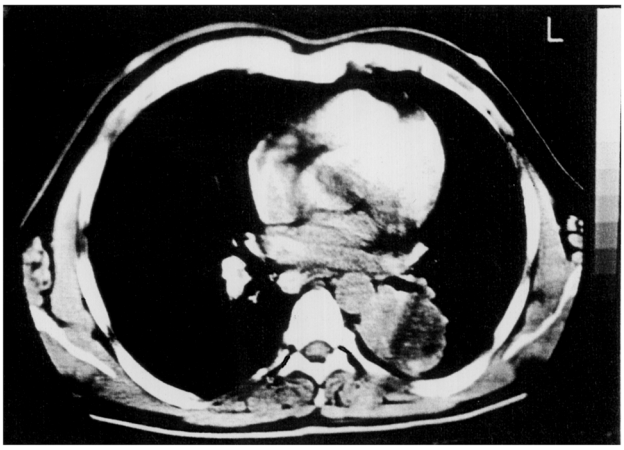


Fig. 1. Thoracic CT scan showing mass.

mainly antracotic, but it was difficult to differentiate them from metastatic tissue. In immunohistochemical studies, the primary tumor cells were staining strongly positive with the melanoma marker (HMB 45) (Fig. 2). The lymph nodes that can not be differentiated from metastatic tissues did not stain with melanoma marker but were positive histological marker (CD 68).

In reference to these results, we decided that the tumor was a malignant melanoma originating from the bronchus epithelium and that there was no hilar lymph node metastasis. Preoperative pathologic diagnosis was non-small cell lung carcinoma with bronchial biopsy. However the histopathologic diagnosis of malignant melanoma may be confused with all malignant epithelial tumors.

A more detailed retrospective research was carried out after the histopathological diagnosis of primary malignant melanoma of the lung. The patient did not have previous cutaneous or mucosal lesion excision performed. The retina, ear, nose, nasopharynx, anal and esophageal mucosa examinations did show nothing but albinoid fundus. The excisional biopsies of the nevi removed from the intergluteal area and from the left side of the nose revealed dermal nevi. The lesion at the left forearm was excised totally and the defect was reconstructed with a full thickness skin graft. Histopathological assessment detected a basal cell carcinoma with clear surgical margins. Consulting with the medical oncology department, we decided to follow the patient without an adjuvant therapy.

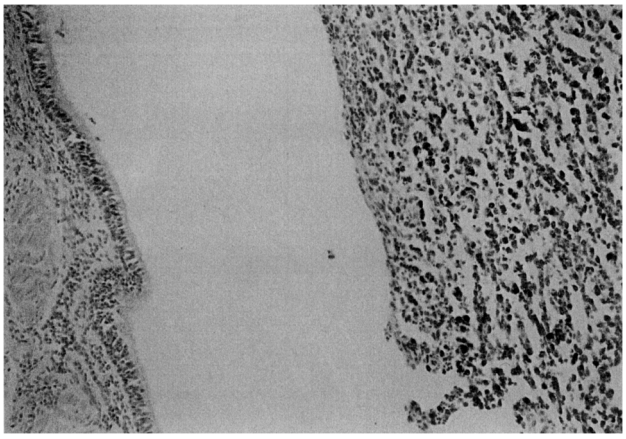


Fig. 2. HMB-45 positive tumor cells (×50).

On follow-up controls, the size and the quality of the nodule in lower lobe of the right lung did not change. In the postoperative 28th month, the patient was admitted to the hospital with weakness, palpitation and epigastric pain. On the examination, iron deficiency anemia and occult blood in stool (+++) were determined. Cranial and thorax CT scans and a whole body bone scintigraphy revealed no abnormalities, but a 10 cm long segment of small bowel with 2 cm thick walls and polipoid lesions in the luminal side were seen on abdominal CT scan. The esophagogastroduodenoscopic and colonoscopic examinations were normal. Small bowel resection was performed and the histopathologic assessment resulted as a malignant melanoma infiltrating the serosa. The patient underwent an interferon treatment for 3 months. In the 37th month the abdominal CT scan showed a 2.5 cm big mass at the right surrenal area. Malignant melanoma was reported after histopathologic examination of the right surrenalectomy material. The patient died of tumor progression in the postoperative 46th month.

2. Discussion

Albinism is an autosomal recessive disorder of melanin synthesis that affects the skin, eyes, and hair. Melanocytes are normally distributed in the eyes and skin; however, normal amounts of melanin are not synthesized [3]. Patients with oculocutaneous albinism are prone to sunlight induced cutaneous malignancy [1,3]. The most frequently encountered tumor is squamous cell carcinoma with basal cell carcinomas seen less frequently [2,3]. Malignant melanoma occurrence is very rare in oculocutaneous albinism [1,3,4]. This low incidence is unexplained [3]. Only 23 cases of malignant melanoma associated with oculocutaneous albinism have been reported in the English literature. Most of these reported lesions were determined in the skin and only one each, ocular, anal and oral mucosal localizations were present [1,3,4].

Because of rarity of association of oculocutaneous albinism and malignant melanoma, further investigation besides histopathologic examination under light microscope was carried out. HMB-45, a specific monoclonal antibody, was also used to demonstrate the melanocytic nature of neoplasms in paraffin- embedded tissue [1,6]. The diagnosis of the tumor excised from the lung was confirmed with detection of HMB-45 positivity.

Primary malignant melanoma of the lung is very rare (Table 1) and carries very rigid diagnostic criters. These include: (1) no previously removed pigmented skin tumor,

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Table 1
Reported cases of primary melanoma[a]

Year	Author	Age (year)	Location	Previous melanoma	Autopsy	Status (time from diagnosis)
1888	Todd et al.	60	ND	ND	+	Dead (1 day)
1888	Tood et al.	55	RLL	ND	+	ND
1916	Kunkel et al.	40	Right hilum	ND	+	Dead (8 months)
1942	Carlucci et al.	48	RML,RLL	0	0	Dead (4 months)
1953	Allen et al.	49	Bronchus	Palate	0	Alive (8 year)
1963	Slam et al.	45	LLL, bronchus	0	+	Dead (6 months)
1964	Reed et al.	66	LUL	Eye	0	Alive (3 year)
1964	Reed et al.	71	LLL	0	0	Alive (10 year)
1965	Rosenberg et al.	46	Main bronchus, trachea	Skin	+	Dead (12 year)
1966	Reid et al.	60	RLL	0	0	Alive (11 year)
1966	Reid et al.	35	Trachea	0	+	Dead (days)
1967	Jensen et al.	61	LUL	0	+	Dead (7 months)
1967	Allen et al.	40	RLL	0	0	ND
1972	Taboada et al.	56	LLL	0	+	Dead (1 year)
1972	Taboada et al.	40	Left hilum	0	0	Alive (3 year)
1980	Weshler et al.	52	Left main bronchus	0	0	Dead (4 months)
1980	Robertson et al.	70	RML	0	+	Dead (9 week)
1981	Gephardt et al.	47	Left main bronchus	0	+	Dead (time of diagnosis)
1984	Carstens et al.	29	RUL	0	+	Dead (1 months)
1984	Cagle et al.	80	RML	0	+	Dead (2 months)
1987	Demeter et al.	56	RUL	0	+	Dead (1 months)
1987	Alghanem et al.	42	LLL	0	0	Alive (2.5 year)
1987	Santos et al.	58	RLL	0	0	Alive (18 months)
1989	Bagwell et al.	62	LUL	0	+	Dead (2 months)
1990	Jennings et al.	34	Left main bronchus	0	0	Alive (19 months)
Present case		41	LLL	0	0	Dead (46 months)

[a] RLL, RML, RUL, right lower, middle, and upper lobes, respectively; LLL, LUL, left lower and upper lobes, respectively; ND, not determined.

preferably no excised skin tumor, (2) no excised ocular tumors, (3) solitary tumor in the surgical specimen from the lung, (4) morphology compatible with a primary tumor, (5) no demonstrable melanomas in other organs at the time of surgery, (6) autopsy without primary malignant melanomas being detected elsewhere, particularly of skin or eyes [5–10]. Taking these criteria into account, our case did not have any excised skin lesions, ocular tumors or enucleation. On histopathological examination, it carried the specification for primary tumor morphology. No melanomas were detected in examination performed prior to the operation. Postoperative cutaneous, retinal, ear, nose, and nasopharynx examinations revealed no finding attributable to melanoma. Histopathological examination of the two totally excised skin lesions; are from the intergluteal and the other from the left nasal regions, were reported as dermal nevi. The lesion at the left forearm was basal cell carcinoma. The lack of abnormal findings in esophagogastroduodenoscopic, colonoscopic, and rectoscopic examinations supported the diagnosis of primary malignant melanoma of the lung. The inability of performing autopsy due to sociocultural reasons was a criteria in the list that could not be fulfilled. Solitary of the tumor was the other criteria that did not match the list. We had four of the six criteria required to make the diagnosis. Thus, it is highly possible that the case reported here is a primary malignant melanoma of the lung. Besides, one of the major problems in making the diagnosis of primary malignant melanoma of the visceral organs is the inevitable possibility of spontaneous regression of the cutaneous lesion.

Considering the clinical progress and survival rate in malignant melanoma, 46 months of the survival is a satisfactory result.

We believe, associations of malignant melanoma with oculocutaneous albinism and especially primary malignant melanomas of the lung render the case value for attention.

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