Melanoma of the Umbilicus: A Patient Report, Precaution in Operative Strategy, and the First Histopathological Review of Published Cases

Shuji Suzuki,* Yuichi Yoshida,* Tatsushi Shiomi,† Shigeto Yanagihara,* Ryoko Kimura* and Osamu Yamamoto*
*Division of Dermatology, Dengatment of Medicine of Sensory and Motor Organs, School of Medicine, Tottori University, Faculty, or

*Division of Dermatology, Department of Medicine of Sensory and Motor Organs, School of Medicine, Tottori University Faculty of Medicine, Yonago 683-8503, Japan and †Division of Organ Pathology, Department of Pathology, School of Medicine, Tottori University Faculty of Medicine, Yonago 683-8503, Japan

ABSTRACT

Umbilical melanoma is extremely rare. Among the past English reports on umbilical melanoma, there are only 8 reports in which histopathology was described in detail, and there has been no report with a review of the histopathology of previously reported cases.

We experienced a case of umbilical melanoma and reviewed previously reported cases including our case. Because of the anatomical location, it is difficult to become aware of the umbilical melanoma unless there are some concomitant symptoms such as discharge or swelling. Even with these symptoms, patients tend to postpone a hospital visit for unknown reasons, resulting in increased risk of tumor growth and metastasis. When performing resection of umbilical melanoma, a portion of the peritoneum should also be removed. Sentinel lymph nodes can be axilla or inguinal lymph nodes. There is a possibility of metastasis to the preoperative abdominal cavity or to nearby skin through hematogenous spread. Preoperative evaluation of tumor spread and postoperative observation are important for umbilical melanoma in order to detect recurrence or metastasis because of its unique anatomical location.

Key words abdominal hernia; peritoneum; prognosis; sentinel lymph node; umbilical melanoma

Primary melanoma arising in the umbilicus is extremely rare. Its anatomical location makes it difficult for the patients to realize the abnormality and challenges surgeons in the treatment strategy. Since Cullen reported the first 3 cases in 1916, 24 cases have been reported in the English literature; however, only 8 cases were reported with sufficient histopathological description. There has been no review of the histopathology of reported cases.

We present a case of melanoma that arose in the umbilicus. We also discuss 7 reports of 8 cases of primary

Corresponding author: Shuji Suzuki, MD, PhD drshuji@yahoo.co.jp Received 2016 March 8 Accepted 2016 April 6 Abbreviation: CT, computed tomography umbilical melanoma with histopathology, locations of sentinel lymph nodes, metastasis, and operative treatment.

PATIENT REPORT

An 83-year-old woman presented to our hospital with an umbilical skin tumor. She first noticed it with exudates and pruritus 2 weeks earlier, saw a local physician, and was referred to us. She had a past medical history of a gastric cancer operation 26 years ago. There was a 12 × 6 mm black nodule arising in the umbilicus without an ulcer on the surface (Figs. 1a and b). No lymph node swelling was palpable on either side of the inguinal and axillary areas.

Biopsy of the nodule revealed a solid proliferation of atypical melanocytes of epithelioid and spindle shapes with tumor thickness exceeding 2 mm. Numerous mitoses were seen and there was no maturation towards the deeper side. Immunostainings against S-100 protein, HMB-45, and Melan-A were all positive. Computed tomography (CT) revealed no metastasis or lymph node swelling. At that time, we diagnosed the nodule as melanoma, T3a (or 4a) N0M0, stage IIA (or IIB). Lymphoscintigraphy with intradermic injections of 99mTc near the nodule showed accumulation of gamma-emitting lymph nodes on both sides of the inguinal areas (Fig. 1c)

Excision with a 2.0 cm margin and sentinel lymph node biopsy of both sides of the inguinal areas were performed after injecting blue dye. The excision line included part of the previous operation scar (Fig. 1d). The peritoneum under the navel was also included within the excision (Fig. 1e). The fascia of each layer and the skin was sutured without obvious tension that might cause the sutures to break at this point.

On the next day after surgery, there was a noticeable swelling of the abdominal wall, and CT of the abdomen revealed an abdominal hernia arising from the umbilical area (Fig. 1f). An emergent radical operation for the hernia using a silicon mesh sheet was performed.

Histopathology of the tumor showed atypical melanocytic proliferation in the dermis invading into the subcutaneous fat layer without maturation (Fig. 1g).

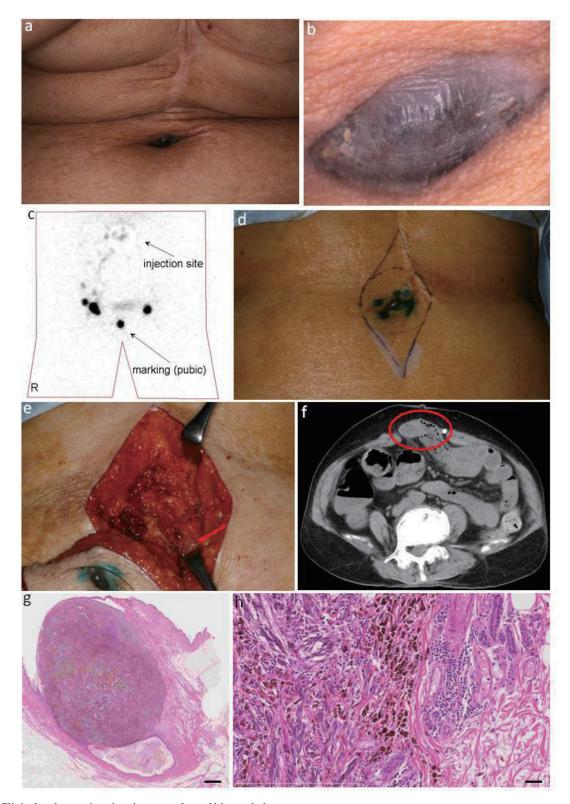


Fig. 1. Clinical and operative view, image study, and histopathology. **a**: A pigmented nodule of 2×1 cm in the umbilicus. **b**: The umbilicus was filled with the nodule. **c**: Lymphoscintigraphy with intradermic injections of 99mTc near the nodule. Accumulation of gamma-emitting lymph nodes on both sides of the inguinal area was visible, being more prominent on the right side. **d**: Incision line included the lower part of the previous operation scar of gastrectomy. **e**: A portion of the peritoneum was removed and a part of the jejunum is exposed (red arrow). **f**: Computed tomography on the first day after tumor resection. A part of the jejunum herniated under the skin (red ellipse). **g**: Atypical melanocyte proliferation in the dermis close to the subcutaneous fat layer (hematoxylin and eosin). Bar = 1 mm. **h**: Some of these cells were in mitosis and no maturation was observed. (hematoxylin and eosin). Bar = 20 μm.

Table 1. Umbilical melanomas

	Age (years old)	Sex	Time between onset/first visit	Size	Clark's level	Breslow tumor thickness	Death/ Alive	Melanoma subtype	Location of SLN	Metastasis	Perito- neum excision
Colonna 1999 ⁴	58	f	Not described	4 cm	V	Not described (>4mm, apparently)	Death, 10 months after resection	Nodular	Not examined	2 satellite skin lesions	+
Colonna 1999 ⁴	30	m	18 mo.	> 10 cm (size of fist)	V	Not described (>4mm, apparently)	Death, 7 months after initial visit	Not described	Not examined	Multiple abdominal and pelvic metastases	
Meine 2003 ⁵	69	f	2 mo.	1.4 × 2.4 cm	Ш	1 mm	Alive 3 y	Superficial spreading	Not examined	-	-/+ (Second surgery)
Campos- Munoz 2007 ⁶	34	f	3 mo.	1.2 cm	III	1.06 mm	Alive 18 mo	Superficial spreading	Left axilla without metastasis	-	+
Mangas 2008 ⁷	63	m	Unknown time	Not described	Ш	0.8 mm	Alive 6 mo	Superficial spreading		-	Not mentioned
Cecchi 2009 ⁸	72	f	4 y	1.5 × 3.0 cm	IV	4.3 mm	Alive 1 y	Superficial spreading + nodular	Both inguinal without metastasis	-	+
Zaccagna 2010 ⁹	60	f	Not described	1.8 × 2.3 cm	IV	2.8 mm	Alive 86 mo	Not described	Left axilla without metastasis	-	+
Song 2013 ¹³	62	m	4 y	2 × 1 cm	IV	3 mm	Alive 36 mo	Not described	Not examined	-	+
Present case	83	f	14 days	1.2 × 0.6 cm	IV	11 mm	Alive 15 mo	Nodular	Both uinal without metastasis	-	+

CT, computed tomography; f, female; m, male; mo, month(s); MRI, magnetic resonance imaging; PET, positron emission tomography; SLN, sentinel lymph node; US, ultrasonography; y, year(s).

Frequent mitoses were observed. Clark's level and Breslow's tumor thickness were IV and 11 mm, respectively (Figs. 1g and h). No melanoma cells were seen in sentinel lymph nodes. The case was diagnosed as melanoma, pT3aN0M0, stage IIA. There have been no signs of recurrence or metastasis of the tumor or of abdominal hernia for 15 months after surgery.

DISCUSSION

Clark et al. described the histopathological level of cutaneous melanoma cell invasion and prognosis in 1969. A review of the histopathology of umbilical melanoma in relation to prognosis was not done until Colonna et al. reported 2 cases of umbilical melanoma in 1999. Many case reports of umbilical melanoma lack histopatho-

logically sufficient evaluation, making it questionable whether the lesions were melanoma or primary lesions. Taking these factors into account, 9 cases including the present case are considered to be histopathologically determined primary umbilical melanoma (Table 1). The age of those patients ranged from 30 to 83 years with an average age of 59 years. The patients included 2 Caucasians, 2 Asians, and 5 patients without ethnical description. Six patients were female.

The present case had the largest described tumor thickness of 11 mm among the 9 cases, but the patient became aware of the tumor only 14 days before the initial visit to a local physician, suggesting that the tumor had existed for a considerably long time. Her abdominal skin sagged and the umbilicus area was not easily visible. She went to hot spas with her daughter about once a month for many years but she first noticed the umbilical nodule when she experienced itching and bleeding after the scratching. There was also a delay in medical care in many of the previously reported cases. Among 5 cases for which there was description about the time between the patient's realization of the lesion and the first medical visit, there were delays of 18 months or more in 3 cases and 2 to 3 months in 2 cases (Table 1). There was a tendency not to visit a physician at an early stage if there were no concomitant symptoms such as discharge or swelling. Sagging skin caused by obesity or aging might be the main reason for not being aware of umbilical lesions. Other patients were aware of the nodules, but they did not seek medical care for a long time for unknown reasons, and the delay resulted in growth and possible metastasis of the tumor.

The long-term prognosis of umbilical melanoma is not clear. Among the histopathologically determined cases, only 2 cases had a follow-up period of 3 years or longer, and the size and thickness of the tumor seem to be important in evaluation of prognosis. Two patients reported died within one year (10 months after resection of the tumor and 7 months after refusing treatment). Both patients had tumors of 4 cm or larger, and, although not specifically described, probably had Breslow tumor thickness far larger than 4 mm judging from the clinical pictures showing large nodules.⁴ Breslow tumor thickness of our case was also very large (11 mm) and there has been no sign of recurrence or metastasis for 15 months after the surgery. It is important to continue to follow up this patient. Four cases were described with histopathological subtypes, superficial spreading in 2 cases, nodular in one case, and both components mixed in the same lesion in one case, and the present case is the second reported case of nodular subtype (Table 1).

The most unique feature of umbilical melanoma is

its anatomical location. Being the embryological remnant of the conduit between the embryo and the placenta, primary melanoma in the umbilicus may metastasize to the abdominal cavity through ligaments derived from the umbilical vein, umbilical arteries, the omphalomesenteric duct, or remnants of the urachus if present.⁹ The tumor may metastasize directly to the adjacent skin through lymphatic or hematogenous spread or to the sentinel lymph nodes. Two cases actually showed metastases: one case had metastasis to adjacent skin and the other case had metastases to multiple abdominal and pelvic organs and to internal thoracic and external iliac lymph nodes.⁴ Among cases in which sentinel lymph nodes were examined, it was found in axillary nodes in 2 cases and in inguinal nodes in 2 cases including the present case. 6, 8, 9 Because of the possibility of metastases through remnant ligaments, 6 of 7 previous reports supported resection of a part of the peritoneum below the umbilicus. ^{4–6, 8, 9, 13} In fact, one case first underwent resection of the fascia level, but remnant melanoma was later found and a second operation was performed one year later to remove the remnant tumor and a portion of the peritoneum.⁵

Regarding the operative treatment approach, all operated cases followed the WHO Melanoma trial for the resection margin, but only one report described augmentation of the abdominal cavity with pre-peritoneal synthetic mesh. 4, 15 We first performed resection of a part of the peritoneum and closed the abdominal cavity, which resulted in postoperative abdominal hernia. Her rectus abdominis may have been brittle due to the previous gastric cancer surgery and her advanced age. Her abdominal cavity should have been reinforced with pre-peritoneal synthetic mesh at the time of resection. Even without pre-surgical fragility of the abdominal wall, resection of the umbilical melanoma with a portion of the peritoneum should be considered with synthetic mesh augmentation.

Umbilical melanoma has a greater chance of tumor growth than does skin melanoma of other areas at the time of the first presentation. Pre-surgical evaluation of tumor spread is necessary, and the resection should be deep enough to include a portion of the peritoneum beneath the lesion.

The authors declare no conflict of interest.

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