Primary Desmoplastic Small Round Cell Tumor of the Submandibular Gland: A Case Report and Literature Review

jiayu zhou
Central South University
Qingling Li
Central South University
baihua luo
Central South University
xiaodan fu
Central South University
chunlin ou
Central South University
xiaomei gao
Central South University
dengyun feng
Central South University
keda yang (YKD820909@163.com)
Central South University

Case Report

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Abstract

Background: Desmoplastic small round cell tumor (DSRCT) is a sporadic, highly malignant tumor with a poor prognosis. The abdomen and pelvis have been reported as the primary localization sites. However, to the best of our knowledge, there are few reports on primary DSRCT in the submandibular gland.

Case presentation: We have reported a case of a 26-year-old Chinese man who presented with a mass in the right submandible. Imaging studies showed a hypoechoic mass in the right mandibular region. Intraoperative pathology revealed that the tumor tissue was composed of small round tumor cells and a dense desmoplastic stroma. On immunostaining, the tumor cells showed markers of epithelial, mesenchymal, myogenic, and neural differentiation. The *EWSR1* gene rearrangement was detected by fluorescence in situ hybridization. Based on the overall morphological features and immunohistochemical findings, a final diagnosis of DSRCT was made. The patient was treated with comprehensive anti-tumor therapy mainly based on radiotherapy and chemotherapy.

Conclusions: DSRCT is a very uncommon disease in which submandibular gland involvement is rare. Considering DSRCT in the differential diagnosis of small round blue cell tumors, even in extraperitoneal locations, is beneficial for a precise diagnosis. The purpose of this report was to describe a cases of DSRCT of submandibular gland and review the literature on the other published cases of DSRCT over the past five years. Full recognition of the clinicopathological features will help to better diagnose this disease.

Background

Desmoplastic small round cell tumor (DSRCT) is an extremely rare and aggressive neoplasm that most commonly affects adolescents and young adults and has a male predominance [1–7]. DSRCT preferentially involves the abdominal and pelvic cavities [30–33]. DSRCT in the pleura, lung, eye, ear, and testis has been reported in only a few cases (< 5%) [3–7], but it is not consistently associated with any specific organ. Clinical symptoms are usually associated with the tumor sites and lack specificity. DSRCT can metastasize in the early stage and quickly recurs despite treatment [18, 36, 38, 39]. Hence, its prognosis is poor. Here, we have presented a rare case of primary DSRCT in the submandibular gland. In addition, we have described and summarized the clinicopathological features of DSRCT and discussed its diagnosis and treatment based on literature review.

Case Presentation

Clinical history

A 26-year-old Chinese man with a chief complaint of a mass in the right submandible for 1 year was admitted to Xiangya Hospital, Central South University, Hunan, China. He had no significant past medical or family history. Routine physical and laboratory examinations were performed. Ultrasonography

revealed a hypoechoic mass measuring approximately 28 mm×18 mm in the right mandibular region, with irregular shape, clear boundary (Fig. 1). Abdominal computer tomography (CT) scan revealed no other lesion elsewhere. There was no evidence of metastasis to the local or distant organs. Hence, lumpectomy was performed under general anesthesia.

Pathology

Histological examination showed sheets, cords, and nests of small round cells separated focally by desmoplastic stroma (Fig. 2a). Under higher magnification, tumor cells showed round to oval hyperchromatic nuclei with an increased nuclear/cytoplasmic ratio and inconspicuous nucleoli. The cytoplasm of the tumor cells was scanty, and the cell borders were indistinct (Fig. 2b). Mitotic activity and individual cell necrosis were common. Immunohistochemical analysis was performed using formalin-fixed paraffin embedded (FFPE) sections of representative tumor blocks and the antibodies listed in Table 1. Immunohistochemical results typically indicated the multi-directional differentiation of tumor cells. The immunohistochemistry results were as follows: Pan cytokeratin(CK-Pan) (+), Epithelial membrane antigen (EMA) (+), vimentin (+), neuron specific enolase (NSE) (+), CD56 (+), chromogranin-A (+), synaptophysin(weakly positive [+/-]), CD99 (+), desmin (+), NKX2.2(-), myogenin (-), S-100(-)and WT1 (-) (Fig. 3). Moreover, the Ki-67 proliferation index was estimated as 50%. The tumor cells were negative for Epstein-Barr virus-encoded small RNA (EBER) on fluorescence in situ hybridization (FISH). And FISH analysis with a break-apart probe proved that the *EWSR1* gene spilt in the neoplastic cells (Fig. 4). However, *EWSR1-WT1* fusion by RT-PCR was not performed due to limited conditions. Based on the above findings and imaging findings, primary lesions in the abdominal cavity and pelvic cavity were excluded, and a final diagnosis of primary DSRCT in the submandibular gland was made.

Table 1
List of immunohistochemical antibodies used in diagnosis

Antibody	Clone	Dilution	Source	Result
AR	EP120	Ready-to-use	Maixin China	+
CD56	MX039	Ready-to-use	Maixin China	+
CD99	O13	Ready-to-use	Maixin China	+
CK-Pan	AE1/AE3	Ready-to-use	Maixin China	+
chromogranin A	193A4C7	Ready-to-use	Baidao China	+
desmin	MX046	Ready-to-use	Maixin China	+
EMA	757F5D6	Ready-to-use	Baidao China	+
FLI1	365H5A6	Ready-to-use	Baidao China	+
IDH1	MX031	Ready-to-use	Maixin China	+
NSE	5E2	Ready-to-use	Zhongshan China	+
synaptophysin	214A4G5	Ready-to-use	Baidao China	Focal+
vimentin	MX034	Ready-to-use	Maixin China	+
CD117	YR145	Ready-to-use	Maixin China	–
CK20	120B1A5	Ready-to-use	Baidao China	–
CK5/6	MX040	Ready-to-use	Maixin China	–
ERG	MXR004	Ready-to-use	Maixin China	–
HHF35	HHF35	Ready-to-use	Zhongshan China	–
Melan-A	A103	Ready-to-use	Maixin China	–
myogenin	F5D	Ready-to-use	Maixin China	–
NKX2.2	EP336	Ready-to-use	Maixin China	–
P63	MX013	Ready-to-use	Maixin China	–
S-100	503F1E9	Ready-to-use	Baidao China	–
TTF-1	MX011	Ready-to-use	Maixin China	–
WT1	MX012	Ready-to-use	Maixin China	–
Ki-67	MX006	Ready-to-use	Maixin China	50%
CK-Pan: Pan cytokeratin; EMA: Epithelial membrane antigen; NSE: Neuron specific enolase;				
+: Positive, –: Negative				

Follow-up

Comprehensive anti-tumor therapy mainly based on chemotherapy and radiotherapy was first proposed. However, synchronous chemotherapy was not performed because of bone marrow suppression. Therefore, cyclophosphamide combined with doxorubicin and vincristine chemotherapy was used for maintenance treatment. The patient is currently alive and well with no evidence of recurrence.

Discussion

Desmoplastic small round cell tumor (DSRCT), which was first described as a specific disease by Gerald and Rosai [8, 9], is a rare and aggressive soft-tissue sarcoma. Generally, DSRCT originates from the serosal surface of the abdominal cavity [30–33], but it can also be found in the lung, eye and salivary gland [3, 43–45, 49–53]. DSRCT has no specific clinical symptoms. Most patients have the initial symptoms of abdominal mass, constipation, ascites, and vomiting [18, 32, 33, 35–38]. It can be accompanied by the manifestations of cachexia, such as fatigue and emaciation. Patients may develop intestinal obstruction, hydronephrosis, and urinary tract irritation owing to tumor compression [15]. In our patient, a right submandibular mass without apparent clinical manifestations was detected incidentally. DSRCT is often widely disseminated throughout the peritoneal cavity, and some patients may present with metastasis to the lymph nodes, liver, and occasionally the lungs [16, 18, 30]. Hence, its prognosis is exceedingly poor. The clinical features of previously published DSRCT cases in the last 5 years are summarized in Supplementary Table 2, Additional File 1.

Imaging examinations of DSRCT lack characteristic features. Ultrasound examination usually shows a lobulated soft tissue mass with an uneven internal echo [17]. Computed tomography (CT) usually reveals single or multiple lobular nodules or lumpy soft tissue masses, with an uneven density of the tumor body and multiple spotted calcifications [32]. The lesions tend to crowd out, surround and invade the surrounding tissues [43]. DSRCT is usually accompanied by flakes of low intensity when there is a necrotic area in the tumor. Enhanced CT presents mild uneven enhancement, and edge enhancement may be observed in some larger masses. Moreover, positron emission tomography (PET)-CT have the potential to monitor residual disease and detect relapse or tumor progression at the early stages [14]. Imaging findings are non-specific, but they can indicate the location, size, and the number of tumors, thereby contributing to biopsy, surgery, and radiotherapy.

The definitive diagnosis of DSRCT is based on typical morphological and immunohistochemical features, especially distinctive molecular characteristics. The pathological and molecular features of previously published cases of DSRCT in the last 5 years are summarized in Supplementary Table 3, Additional File 2. Histologically, the tumor tissue consists of small round cells and peripheral desmoplastic stroma, which can be accompanied by cystic degeneration and hemorrhagic necrosis. The tumor had a variegated histology revealing pseudopapillary architecture, rhabdoid, clear or pleomorphic cells in addition to typical small round cell morphology. Moreover, the tumor can have intermittent areas of primitive tubules or rosette-like structures [18]. Immunohistochemically, tumor cells show a pattern of

multi-phenotypic differentiation [30, 39–41]. This multiple antigen expression profile is a characteristic of DSRCT and can be used to distinguish DSRCT from the other histologically related small round cell tumors. Further, para nuclear dot-like desmin positivity has important diagnostic significance. However, in our case, immunohistochemical staining showed a diffuse perinuclear staining pattern with desmin, but characteristic dot positivity was not prominent. Almost all cases of DSRCT are positive for WT1 and show cytoplasmic and paranuclear staining. Although the immunohistochemical analysis of classic cases of DSRCT tend reveal WT1 positivity, N- and C-terminals may be useful as a form of “molecular immunohistochemistry” to identify the EWS–WT1 transcript as the immunostaining pattern may be altered by variant transcripts and WT1 immunostaining may be negative (as in our case) [46–48]. To establish a DSRCT diagnosis, the interpretation of WT1 immunostaining requires knowledge of antibody target epitopes and correlations with clinical, morphological, and molecular findings.

In the present case, the tumor was composed of nests of small to medium-sized cells, which might be misdiagnosed as small cell carcinoma. Small cell carcinoma can also show the immunoreactivity for epithelial and neuroendocrine markers. But in our case, immunohistochemical analysis of the co-expression of EMA, vimentin and desmin by tumor cells strongly support a diagnosis of DSRCT. DSRCT should also be distinguished from other carcinomas such as malignant melanoma, malignant lymphoma and metastatic neuroblastoma. However, the current case was negative for Melan-A and S100, which made malignant melanoma unlikely. And the positivity for epithelial markers helped to rule out the possibility of malignant lymphoma, which often involves lymph nodes, bone marrow and peripheral blood. In addition, stroma of massive nerve fiber network is a characteristic feature in neuroblastoma, which might be a diagnostic clue. Due to the histological features of small round cells in the present tumor, it must be distinguished from other small round cell tumors such as rhabdomyosarcoma, primitive neuroectodermal tumor (PNET) and Ewing sarcoma (EWS). Rhabdomyosarcoma is more common in children; the tumor cells are commonly positive for myogenic markers (such as MYOD1, myogenin), but negative for epithelial and neuroendocrine markers. Morphologically, DSRCT and PNET can revealed chrysanthemum-like structure, and both of them have positive expression of CD99 and NSE. In our case, immunohistochemical results show the positivity for desmin and epithelial markers, which favors a diagnosis of DSRCT over that of PNET. EWS shares histological and immunophenotypic similarities with DSRCT. However, the survival rate of patients with DSRCT is significantly lower than that of EWS patients, which indicates that DSRCT and EWS have different biological backgrounds. EWS mainly occurs in children and is common in bones, EWS involving soft tissue is rare. EWS can also be positive for cytokeratin, desmin, CD99, FLI-1 and neuroendocrine markers, which may cause confusion. The diffuse membranous positivity of CD99 is typical of EWS, but in our case, nonspecific cytoplasmic positivity for CD99 is one of the features of DSRCT. Furthermore, in our case, negative of NKX2.2 is an important clue to distinguish EWS. Both EWS and DSRCT harbor *EWSR1* rearrangements, the break-apart FISH assay for *EWSR1* will not be helpful in the differential diagnosis between them. But characteristic translocation of EWS involves *EWSR1* and the ETS family of transcription factors, not *WT1.* Convincingly, documentation of *EWSR1-WT1* fusion is the “gold standard” for the diagnosis of DSRCT [35, 39, 40]. It was not performed in our case due to limited conditions. Taken together, combined with the tumor location and

morphological features, as well as a distinctive pattern of multi-phenotypic differentiation on immunohistochemistry, we prefer the diagnosis of DSRCT. Like DSRCT, both myoepithelial carcinoma and synovial sarcoma are also multiphenotypic and expresses multilineage markers. Myoepithelial carcinoma expresses cytokeratin and myogenic markers such as myogenin, smooth muscle actin (SMA) and HHF35. The case in this study is positive for desmin and negative for all other myogenic markers, P63 and CK5/6, strongly favors a diagnosis of DSRCT. Synovial sarcoma mainly occurs in the extremities. It expresses epithelial and mesenchymal markers, but desmin positivity is uncommon, which can help in the differentiation.

DSRCT is distinguished by the t (11;22) (p13; q12) chromosomal translocation involving a fusion between the transcriptional activating domain of *EWSR1* and the *WT1* gene [19–21]. Studies have also suggested that the *EWSR1-WT1* fusion protein can induce the expression of PDGFA. PDGFA can induce the growth and proliferation of fibroblasts and the production of collagenous stroma, which may explain the characteristic reactive fibrosis of DSRCT [22]. Downstream activation of *EWSR1-WT1* gene fusion includes signaling pathways of vascular endothelial growth factor (VEGF), IL2RB, and insulin growth factor (IGF)-1 [23–25]. A better understanding of the effects of these target genes will provide avenues for future treatment.

Despite multimodality treatment, DSRCT is highly aggressive and has a poor prognosis. The overall survival in patients is < 3–5 years after diagnosis, and the 5-year survival rate is < 20% [18, 26, 39, 48]. There is no standardized approach for the treatment of this malignant disease. Effective cytoreduction combined with comprehensive therapies, as the best treatment strategy presented in most studies, may prolong patient survival [27, 37–41]. With the in-depth analysis of molecular genetics of DSRCT, targeted therapy, immunotherapy and other methods have been used for the treatment of DSRCT in recent years [28, 29].

Conclusions

In summary, DSRCT is a poorly understood malignant tumor with characteristic morphology, immunophenotype, and cytogenetic features. The disease does not present with specific clinical signs or symptoms. PET-CT may help diagnose recurrent disease at an earlier stage. The *EWSR1-WT1* fusion by RT-PCR is the gold standard for the diagnosis of DSRCT. When it is not feasible, definitive diagnosis mainly depends on a comprehensive analysis of histological and immunohistochemical studies. The submandibular gland is an unusual site for DSRCT, suggesting that the tumor may not have a site-specific predilection.The pathologist and the clinician have to be aware of its possible occurrence in extra-peritoneal regions. To better diagnosis this rare and intriguing disease, further studies are needed in future studies.

Abbreviations

DSRCT: Desmoplastic small round cell tumor

CT: Computed tomography

PET: Positron emission tomography

FFPE: Formalin-fixed paraffin embedded

CK-Pan: Pan cytokeratin

EMA: Epithelial membrane antigen

NSE: Neuron specific enolase

EBER: Epstein-Barr virus-encoded small RNA

FISH: Fluorescence in situ hybridization

RT-PCR: Reverse transcription-polymerase chain reaction

PNET: Primitive neuroectodermal tumor

EWS: Ewing sarcoma

Declarations

Authors' contributions

Jiayu Zhou is a major contributor in writing the manuscript and compiling figures. Qingling Li, Baihua Luo and Xiaodan Fu contributed to the design and format of figures and tables. Chunlin Ou and Xiaomei Gao helped revising the manuscript. Deyun Feng and Keda Yang confirmed the pathological analysis. Keda Yang designed and organized the study. All authors read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this case are included within the article.

Ethics approval and consent to participate

This case study was approved by the Institutional Ethics Committee of Xiangya Hospital Central South University, Hunan Province, China.

Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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Figures

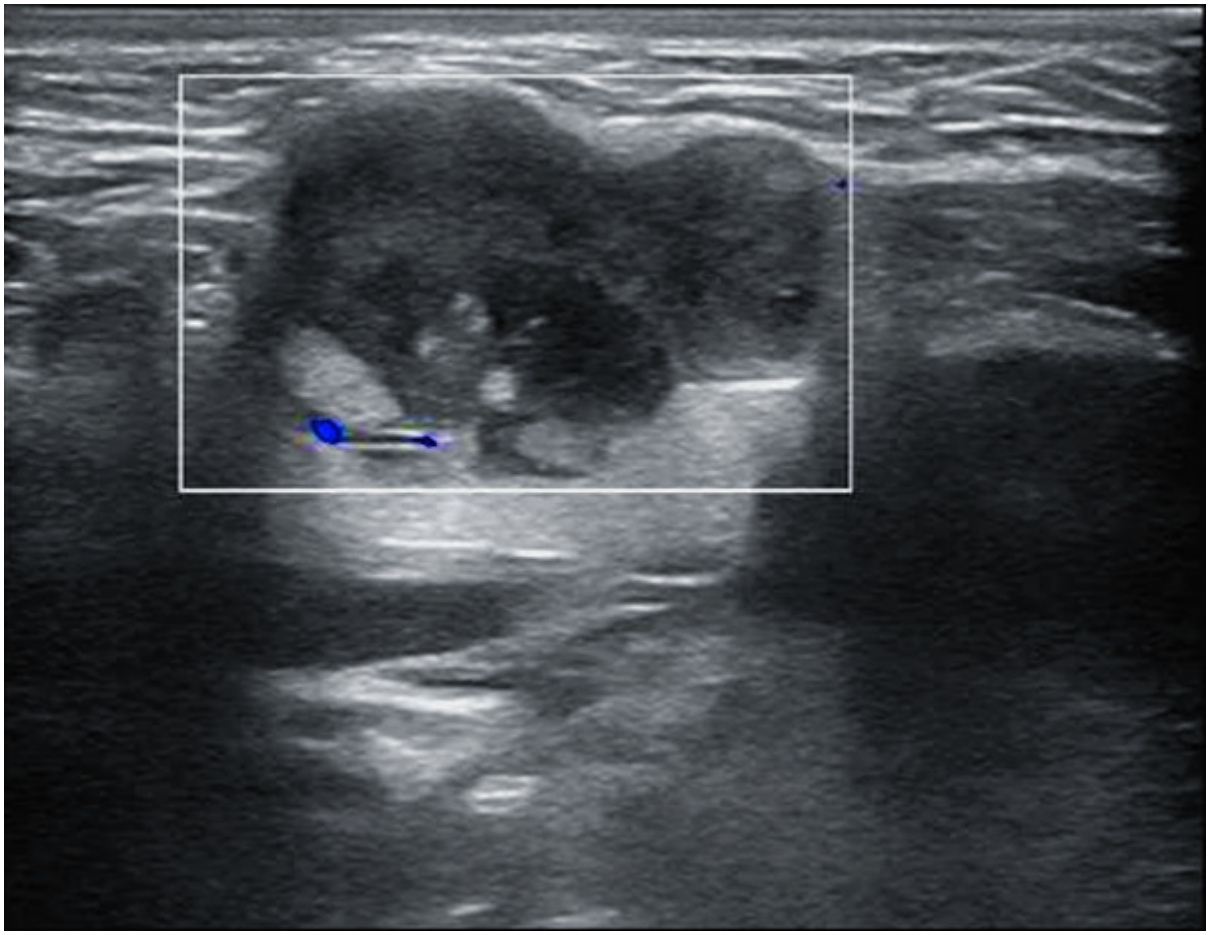

Figure 1

Cervical US: A hypoechoic, well-defined mass measuring 28 mm ×18 mm located in the right submandibular region; no obvious blood flow signal was observed in the lesion.

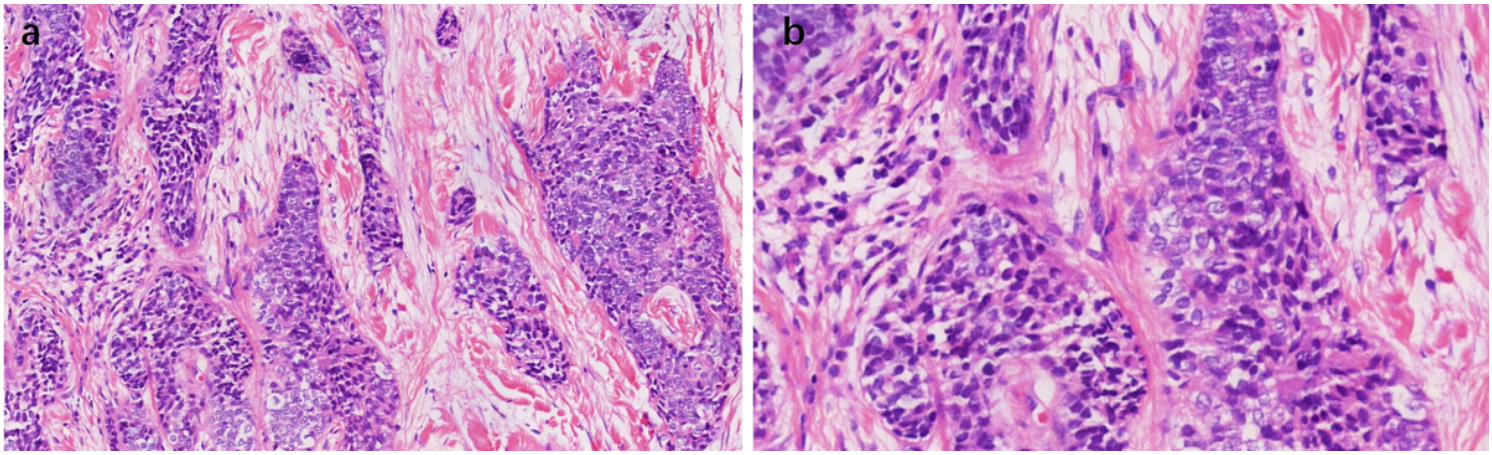

Figure 2

(a) Tumor cells were irregular sheet-like and nest-like distribution, surrounded by proliferative fibrous stroma. (H&E, magnification ×200). (b) The tumor cells are small round or oval, with few cytoplasm, unclear cell boundaries, round or oval hyperchromatic nuclei and unclear nucleoli (H&E, magnification ×400).

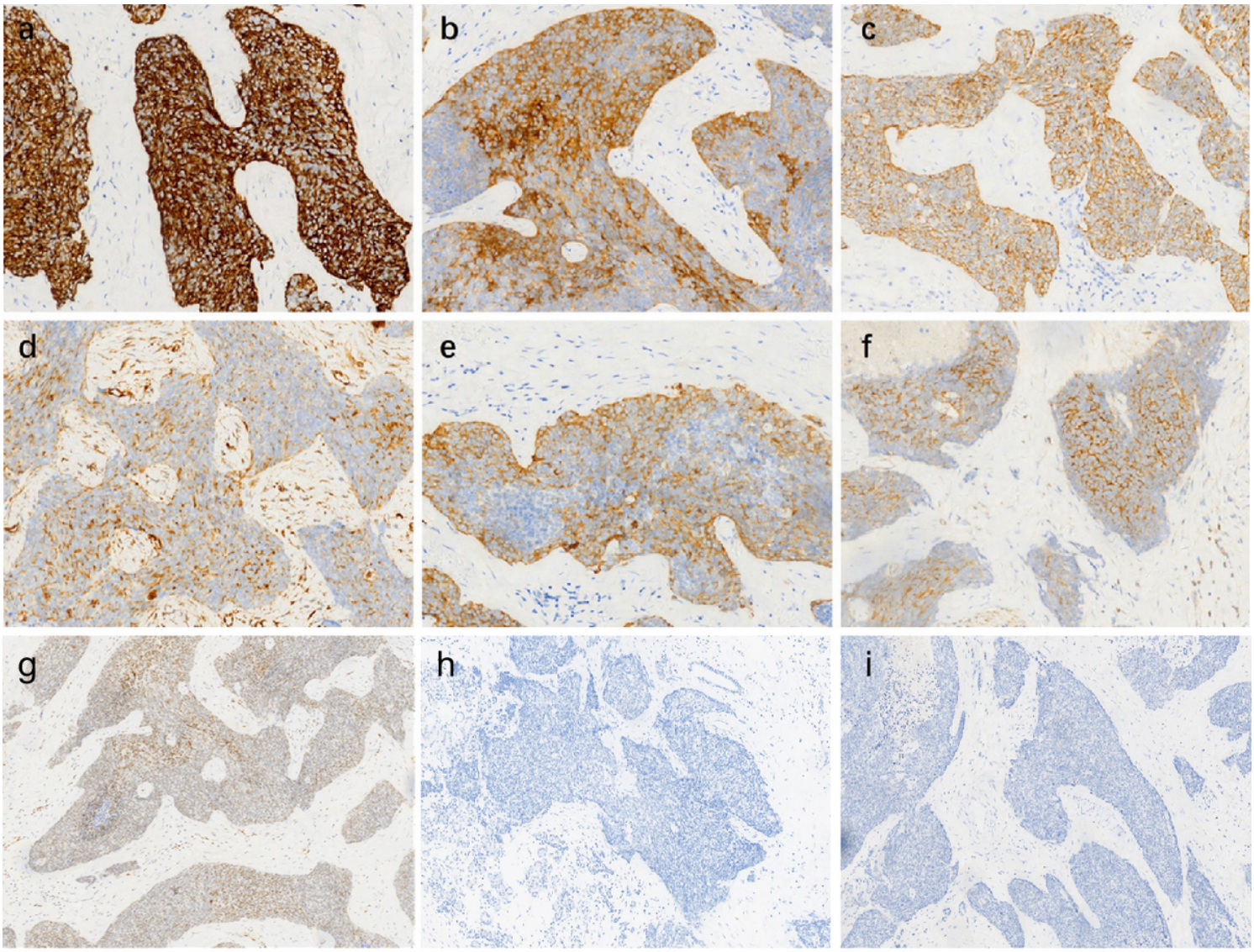

Figure 3

Neoplastic cells are positive for desmin (a), neuron specific enolase (NSE) (b), CK-Pan (c), vimentin (d), synaptophysin (e), CD99 (f) and FLI-1 (g), but negative for NKX2.2 (h) and WT-1 (i).

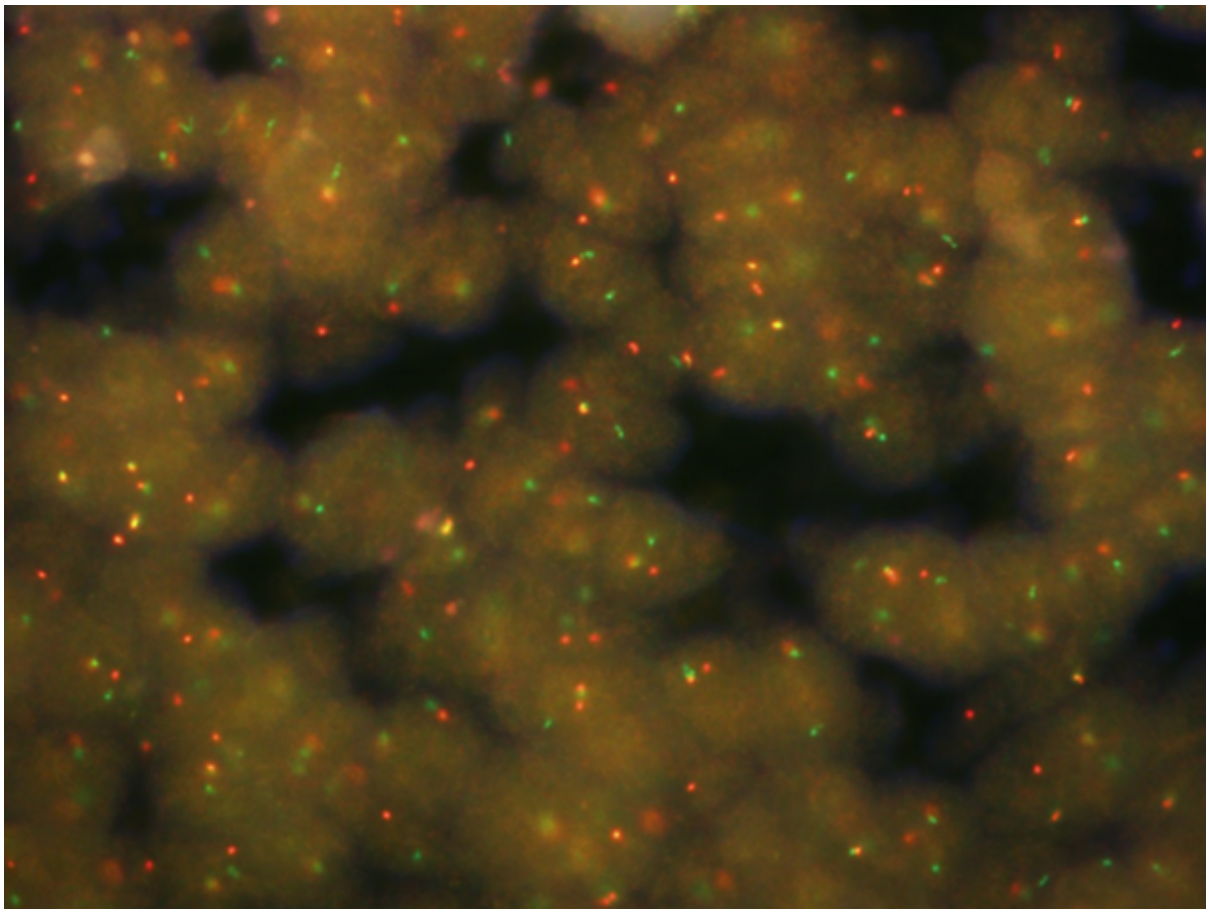

Figure 4

Dual Color Break Apart specific locus FISH probe targeting EWSR1 gene; green and red signals mark the 5' and 3' ends of the gene respectively.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Additionalfile1.doc
- Additionalfile2.doc