Cavitary Retinoblastoma: Ultrasonographic and Fluorescein Angiographic Findings in 3 Cases

Retinoblastoma is an important, life-threatening intraocular malignancy of childhood and it is recognized by classic ophthalmoscopic features. Generally, retinoblastoma appears as a dome-shaped, solid white retinal mass with prominent intrinsic and feeder vessels. Rarely, it manifests with ophthalmoscopically visible cavities within the tumor. The few previous reports on cavitary retinoblastoma have described its relative chemoresistant and radioresistant features. In this report, we describe the ultrasonographic and fluorescein angiographic features of cavitary retinoblastoma.

Report of Cases. The 3 cases are listed in the Table and described in this section.

Case 1. A 24-month-old girl with bilateral retinoblastoma manifested a retinoblastoma in her right eye classified as group C according to the International Classification of Retinoblastoma. The tumor measured 9.0 × 8.0 mm in diameter and 5.0 mm in thickness and displayed 4 cavities, each measuring approximately 2.0 mm or less in diameter (Figure 1). On A-scan ultrasonography, the retinoblastoma showed high internal reflectivity and the cavities showed low internal reflectivity and B-scan showed scattered intrinsic calcification with no subretinal fluid and the cavities showed subtle echolucency with well-defined borders. On fluorescein angiography, the tumor was hyperfluorescent starting from the venous phase and the cavities showed hypofluorescence throughout all phases. Following 6 cycles of chemoreduction (CRD) and thermotherapy (TTT), the tumor showed slight but stable regression to 8.0 mm in diameter and 4.0 mm in thickness.

Case 2. A 9-month-old boy with bilateral sporadic retinoblastoma was found to have cavitary retinoblastoma in the left eye (Figure 2). The tumor basal diameter was 14.0 × 13.0 mm and tumor thickness was 11.0 mm. The cavity was approximately 3.0 × 3.0 mm. The tumor displayed high internal reflectivity on A-scan and prominent calcification and related subretinal fluid on B-scan ultrasonography. The cavity displayed low internal reflectivity on A-scan and was echolucent on B-scan ultrasonography (Figure 2). The tumor was hyperfluorescent on fluorescein angiography and the cavity was nonfluorescent (Figure 2). Following 6 cycles of CRD and TTT, the tumor regressed to a partially calcified remnant measuring 13.0 mm in basal diameter and 4.5 mm in thickness.

Case 3. A 15-month-old girl with bilateral sporadic retinoblastoma displayed retinoblastoma in her left eye manifesting 2 cavities (Figure 3). The tumor basal diameter was 15.0 × 15.0 mm and thickness was 7.0 mm. Each cavity was 3.0 mm in diameter. On A-scan ultrasonography, the tumor showed high internal reflectivity while the cavities

| Table. Clinical and Imaging Features of 3 Cases of Cavitary Retinoblastoma |
|-----------------------------|-----------------|-----------------|-----------------|
| Features                    | Case 1          | Case 2          | Case 3          |
| Retinoblastoma heredity     | Bilateral sporadic | Bilateral sporadic | Bilateral sporadic |
| Laterality                  |                 |                 |                 |
| Classification and management |                 |                 |                 |
| Right eye                   | Group C: chemoreduction + TTT | Group E: chemoreduction + EBRT | Group D: chemoreduction + TTT |
| Left eye                    | Group E: enucleation | Group B: chemoreduction + TTT | Left eye |
| Eye with cavitary retinoblastoma |                 |                 |                 |
| Tumor base, mm              | 9.0 × 8.0       | 14.0 × 13.0     | 15.0 × 15.0     |
| Tumor thickness, mm         | 5.0             | 11.0            | 7.0             |
| No. of cavities per tumor   | 4               | 1               | 2               |
| Largest cavity diameter, mm | 2.5 × 2.5       | 3.0 × 3.0       | 3.5 × 3.0       |
| Largest cavity, % of entire mass | 4.3             | 1.3             | 2.0             |
| A-scan ultrasonography      |                 |                 |                 |
| Tumor                       | High reflectivity | High reflectivity | High reflectivity |
| Cavity                      | Low reflectivity | Low reflectivity | Low reflectivity |
| B-scan ultrasonography      |                 |                 |                 |
| Tumor                       | Mild calcification | Mild calcification | Mild calcification |
| Cavity                      | Circumscribed, echolucent | Circumscribed, echolucent | Circumscribed, echolucent |
| Fluorescein angiography     |                 |                 |                 |
| Tumor                       | Hyperfluorescent | Hyperfluorescent | Hyperfluorescent |
| Cavity                      | Hypofluorescent | Hypofluorescent | Hypofluorescent |

Abbreviations: EBRT, external beam radiation therapy; TTT, thermotherapy.

Classification according to the International Classification of Retinoblastoma.
showed low reflectivity (Figure 3). On B-scan ultrasonography, the tumor showed mild calcification with no subretinal fluid, and both cavities were echolucent (Figure 3). On fluorescein angiography, the deeper cavity was not visible while the superficial cavity was hypofluorescent (Figure 3). After 6 cycles of CRD and TTT, the retinoblastoma showed a partially calcified regressed remnant with only slight clinical response measuring 8.0 mm in basal diameter and 2.0 mm in thickness, with persistent visibility of the cavities (Figure 3).

Comment. In 1952, Samuels and Fuchs provided a diagram of a globe with retinoblastoma that showed a small well-defined cyst near its apex. They suspected that tumor liquefaction was the origin of the cyst. Although terms of cyst and cystic retinoblastoma have been used previously by some authors, because of the histopathologic absence of definite lining cells, Mashayekhi and coworkers offered the terms cavity and cavitary retinoblastoma for reference to this entity.

Retinoblastoma usually demonstrates an initial dramatic response to chemotherapy with a mean tumor reduction of 35% to 50% after 2 cycles of chemotherapy. Lack of response to therapy is suggestive of a more benign variant of retinoblastoma like retinoma or retinocytoma, otherwise known as spontaneously arrested or regressed retinoblastoma. Additionally, lack of response has been observed with cavitary retinoblastoma, believed to be due to the presence of features of retinoma and retinocytoma within the mass. Well-differentiated retinoblastoma, like cavitary retinoblastoma, shows only slight response to chemotherapy, often with regression to a noncalcified remnant. All 3 of our cases showed little response to CRD. Two of our cases (case 2 and case 3) have been included in a previous analysis on tumor regression following CRD. In this report, we elucidate the diagnostic testing results of eyes with cavitary retinoblastoma. We found that the cavities were all echolucent on ocular ultrasonography and showed hypofluorescence, in contrast to the bright hyperfluorescence of the tumor, on fluorescein angiography. These fluorescein angiographic and ultrasonographic features provide clues to the diagnosis of cavitary retinoblastoma and predict a less dramatic response to CRD.

The differential diagnosis of cavitary retinoblastoma includes cystic astrocytoma, atypical granuloma, localized retinoschisis, and cavitary amelanotic melanoma with retinal invasion. The most important concern with cavitary retinoblastoma is its poor response to chemotherapy and radiotherapy, where it can be confused with resistant viable reti-
noblasm or recurrent retinoblas-
toma. The long-term stability of
treated cavitary retinoblastoma as-
sists in confirming its regressed state.

In summary, cavitary retinoblas-
toma can be recognized by oph-
thalmoscopy. In this report, the
ultrasonography and fluorescein
angiography findings are illus-
trated and they can assist in sup-
porting the diagnosis. Poor visible
response to chemotherapy should
not be deemed as treatment failure
but should be anticipated, based on
the initial clinical features of cavi-
tary retinoblastoma, and the eye
should be cautiously observed.

Melis Palamar, MD
Cesare Pirondini, MD
Carol L. Shields, MD
Jerry A. Shields, MD

Correspondence: Dr C. L. Shields,
Ocular Oncology Service, Ste 1440,
Wills Eye Institute, 840 Walnut St,
Philadelphia, PA 19107 (carol.
shields@shieldsoncology.com).

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